ESTHIOMÈNE OR LUPUS VULVAE.

A Historical, Pathological and Clinical Study.

(With one colour sketch and 10 microphotographs.)

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CHAPTER I.

Introductory Notes.

In the year 1849 Huguier published his remarkable essay entitled: "de l'Esthiomène de la Vulve et du Périnée." He derived the name from the Greek word ἐσθίομενος which signifies to eat away. Previous to the appearance of this essay every variety of vulvar ulceration had been described under the term of "lupus" irrespective of the cause and nature of the ulceration described. After the publication of Huguier's essay, numerous French and English authors adopted the term of esthiomène and many /

many of them retained the term of lupus concurrently. It is obvious that many writers on the subject understood by both terms merely a process of "ulcerative destruction" and did not intend to point to any definite causation by the choice of name.

Definitions.

Huguier's own definition of the condition is as follows:-

"Cette maladie chronique qui tient le milieu entre l'éléphantiase des Arabes, la syphilis, le cancer et la scrofule, quand elle n'est pas essentiellement de cette dernière nature, est charactérisée par la teinte plombée et violacée des parties, leur déformation, leur induration et épaississement, leur ulcération, destruction hypertrophique et infiltration simultanées, de telle sorte que les orifices et les cavaux qu'offre la region vulvo-anal peuvent être en même temps ulcérés agrandis et retrécis, les sillons, les replis cutanés et mugueux plus developés épaissis et le siège d'ulcérations et de cicatrices plus ou moins étendues et profondes, sans douleurs ni élancements sans menacer directement la vie ni même porter de long temps une atteinte profonde à la constitution."

From this description it is obvious that the author of the essay on Esthiomène has no view as to the nature and causation of the condition he describes. But immediately after his publication some /

some authors maintained that Huguier created a "new disease", and others held the view that since Huguier retained the term of lupus he meant to imply a tuberculous ulceration. All that Huguier did in reality was to create a new name under which he described a series of cases of vulvar ulceration which to us appear undoubtedly syphilitic. He did not commit himself to any theory of causation but limited himself to a masterly description of a number of ulcerations occurring in the region of the Vulva and anus.

From the first the interest in esthiomène has centred around the theories of causation and a long controversy both in English and French Literature has arisen from faulty observation and insufficient microscopic examination. In our days almost every case of "Esthiomène" can be definitely diagnosed by the aid of the microscope and we have no doubt as to the real causes of "esthiomenic ulceration."

Guibout in an article in "l'Union Médicale" 1849 describes esthiomène as "une affection chronique qui donne aux chairs une teinte bleuâtre, indure les parties à mesure qu'elle les détruit et ne s'accompagne /

s'accompagne de symptômes généraux et n'attaque que des adultes."

A few years later A. Boursier in his Précis de Gynécologie (reprinted in 1903) defined his condition in these terms:-

"L'Esthiomène de la Vulve est une ulcération chronique due ordinairement à des causes banales et se compliquant d'un élément hypertrophique résultant d'un processus éléphantiasique."

Among the English writers Mathews Duncan 1885 gave the following definition of Esthiomène:-

This disease gets its name from the destruction which it causes by ulceration, and although many cases are marked more by hypertrophy or growth than by destruction, the quality of "exedens" is very common and strikingly peculiar as well as most injurious and intractable."

A general survey of all the definitions of Esthiomène put forward by French and English authors on the subject makes it at once clear that they all agreed in considering it a chronic ulcerative condition of the Vulva, invariably accompanied by hypertrophy and induration of the surrounding parts; that ulceration /

ulceration and hypertrophy go on side by side varying in degree; that ulceration may be slight (lupus minimus of Mathews Duncan) or it may be extensive and show a tendency to burrow (lupus Exedens or lupus maximus); thatthe etiology is uncertain and the morbid anatomy indefinite; that the terminology is usually fatal where ulceration is severe but that the condition is amenable to treatment where hypertrophy is the main feature.

Terminology.

Esthiomène has been frequently described under the term of lupus to which all varieties of descriptive adjectives have been added, such as:- lupus hypertrophicus, lupus serpiginosus, lupus prominens, lupus perforans etc. according to the main feature presented by the case.

The term lupus so used did not imply tuberculosis. Another name frequently met with is "Herpes", Herpes exedens and sometimes "herpes esthiomène".

German authors in contradistinction to the French and British writers rarely adopted the name of "lupus" and the term esthiomène is only introduced when the French writers are quoted by the Germans. Virchow introduced the term:- "Ulcus Rodens Vulvae" which was taken up by Veit and other German gynaecologists, while F. Koch adopts the term "Ulcus chronicum elephantiasticum".

Recent authors both Continental and British have used such terms as:- "lupoid ulcer" and genito-anal scleroma" when describing "esthiomenic" ulcerations.

The large variety of names points to two important /

important facts: firstly, that the writers did not describe one definite type of ulcer with hypertrophy but absolutely every kind and type of vulvar ulceration, and secondly that the nature of esthiomenic ulceration was quite unknown and only tentatively hinted at in the particular terms used. Many authors did not intend to commit themselves to any theory of causation and they chose the term "ulcus"; others understood only "ulcerative destruction" by the name lupus or esthiomène; a smaller group understood tuberculosis by lupus and a "special disease" by the term of esthiomène.

The theories of causation will be discussed in a subsequent chapter.

CHAPTER II.

Historical Outline.

The first work known on the subject of "Esthiomène or lupus Vulvae" is Huguier's masterly essay entitled "De l'Esthiomène de la Vulve et du Perinée", which he presented to the French Academy of Medicine and which was published in "the Mémoires de l'Académie de Médicine" in 1849. Huguier created the term of esthiomène from the Greek word ἐσθίομενος to eat away with the intention of indicating the ulcerating and sometimes burrowing nature of the condition and excluding any theory as to the nature of the disease. As already indicated in the "introductory notes" Huguier did not exclude the term lupus from his publication, and thus gave rise to long controversies on the part of those who interpreted him to be an advocate of the theory that esthiomène or lupus is a tuberculous condition and others who maintained that he meant a disease apart, "sui generis". We have already seen that Huguier held no views with regard to the aetiology of esthiomène. He expressly retained the name lupus because he saw a definite resemblance between cases of /

of esthiomenic ulceration and Biet's the French dermatologist's classification of cases of Lupus:- the latter divided them into:-

1. Lupus which destroys on the surface.
2. Lupus which destroys the deep structures.
3. Lupus with hypertrophy.

These three varieties occur undoubtedly in the condition that Huguier has termed esthiomène, but he did not adopt the name of lupus in order to draw an inference as to similarity of aetiology and morbidity but merely because he saw a clinical resemblance. He classified his esthiomenic cases on the same lines as follows:-

1. Superficial ambulant and serpiginous variety:-
   a. Erythematous.
   b. Superficial, tuberculated, serpiginous.
2. Perforating Variety.
3. Hypertrophic Variety:-
   a. Hypertrophic Vegetating.
   b. Hypertrophic, oedematous and elephantiasic.

His classification and nomenclature have been adopted by a large number of French authors and by a few British ones.

It /

It would be difficult to find anywhere in medical literature a more masterly description of morbid changes than Huguier presented us with when he appended to his essay the clinical study of six cases of esthiomène. It is not surprising that to this day his work has remained one of the best known and most frequently quoted of French essays. In spite of his avoidance of any discussion regarding the nature of Esthiomène, necessitated by the lack of knowledge of the causation of Syphilis and lupus, and the insufficient histological methods of his time, Huguier's essay is one of primary importance in the study of the ulcerative conditions of the vulvo-anal region to which all students of ulcerative lesions connected with general constitutional diseases should refer.

In the same year in which Huguier published his thesis, Guibout wrote an essay in "l'Union Médicale" entitled: "Des diverses affections non-vénériennes des organes genitaux-urinaires, chez la femme qu'on est exposé à considérer comme vénériennes." Amongst a number of varieties of non-specific ulcerations of the vulvo-anal region Guibout describes those types which /

which he unites under the heading of "Esthiomène or Lupus of the Vulva." The author attempts to distinguish these types from

a. Syphilis

b. Cancer

c. Elephantiasis Arabum.

His treatise deals mainly with the aetiology of esthiomène and with the differential diagnosis between esthiomène and the conditions mentioned. He concludes his observations by stating that lupus of the face and lupus of the vulva are one and the same disease: "il y'a réellement à la Vulve qu'à la face un esthiomène." He considers that lupus whether it occurs in the face or on the Vulva is a very definite disease of unknown etiology. He does not theorise as to its possible causation.

Bernutz in the "Archives de Tocologie" (1874) published an essay on esthiomène in which he follows Guibout's example in upholding the identity between lupus of the face and lupus of the vulva. He differs from Guibout, however, in recognising the "scrofulous" by which he means tuberculous, nature of lupus. Both cases described in his essay were scrofulous. One had /

had a history of "scrofula," the other suffered from advanced phthisis of which the patient died. Bernutz is the first author on esthiomène who forwarded the view that lupus whether it appeared on the face or on the vulva was a tuberculous ulceration and who definitely associated the ulceration termed esthiomène with constitutional tuberculosis. His views were-accepted by a series of French authors until Eugène Deschamps in 1885 issued his "Étude sur quelques ulcérations non-vénériennes de la Vulve et du Vagin" in the Archives de Tocologie." He discarded the traditional views that esthiomène is lupus in the same sense as lupus of the face and that it is part of a general tuberculous condition.

In his essay Deschamps deals mainly with the theories of causation of esthiomène passing in rapid review all the chief theses that had up to that time appeared in the French publications. Deschamps' view was that esthiomène was not due to any one particular cause but to different causes such as cancer syphilis and tuberculosis. He considered that the etiology in each case of esthiomenic ulceration could only be determined by the response to /

to treatment and the course of the disease.

Another "thèse de Paris" on the subject of Esthiomène was written by Figuet in 1876, it is entitled: "Essay sur l'Esthiomène de la region Vulvo-anale, étude sur quelques ulcérations non-vénériennes de la Vulve et du Vagin." Here again the main part of the work deals with the aetiology and the author's observations lead him to consider like Bernutz that esthiomène is a manifestation of scrofula in the sense of tuberculosis. "L'Esthiomène est une affection strumeuse qui coexiste avec d'autres scrofulides cutanées, qui peut être une scrofulide fixe primitive et qui est en un tiers de cas accompagnée de tuberculeuse pulmonaire."

The next contribution in point of time to the literature of esthiomène appeared in 1887 in New York in the shape of a monograph published by Isaac Taylor M.D. in the "Gynaecological transactions of Philadelphia" entitled: "On Lupus or Esthiomène of the Vulvo-Anal Region." The author holds that patients afflicted with Esthiomène are usually free from "scrofula", and he therefore uses the term lupus in the sense only of destruction. Isaac Taylor is /

is the first to incline to a view that esthiomène or lupus might from several peculiarities which appartain to this affection be considered a disease "sui-generis." Like Huguier he forwards no view as to its possible causative factors but is inclined to think it a "local disease grafted on a constitutional one such as cancer syphilis or tuberculosis."

The first British author who contributed a valuable paper to the literature on esthiomène was Angus Macdonald who in 1883 published three cases in the Obstetrical Transactions Edinburgh under the name of "Lupus of the Vulvo-anal Region."

In the dissertation incidental to the report of the three cases he refers to Huguier's Essay (1849) and adopts his classification. Regarding his view on the nature of the condition he thinks it is "lupus" but not in sense of "scrofula" but in the same sense as lupus of the face; "there is no essential difference between lupus of the vulva and lupus on any other part of the body." "Various attempts have been made to connect lupus with certain cachectic conditions such as syphilis or scrofula; the latter view is especially common among French authors. In considering /

considering the three cases which form the subject matter of this paper neither the taint of syphilis nor the slightest evidence of any strumous or tubercular condition could be detected. It would appear therefore that lupus must be looked upon as a local not a general affection its origin having to be sought for in some local irritation." He means therefore that "lupus" implies merely an "ulceration" and there is no connection in his mind between scrofula or tuberculosis,(which he considers to be a general constitutional disease)and "lupus" of the face or perineum which to him are merely local ulcers of unknown origin.

Mathews Duncan published two valuable monographs on Lupus or Esthiomène which appeared in 1884 and 1885. The first "On Lupus of the Pudendum"was printed in the Medical Times Vol. II. 1884. The author adheres to the term lupus because, as he explains, he sees a resemblance between lupus of any part of the body, and lupus of the perineum, but he does not admit that they are "pathologically the same as is generally believed."

He distinguishes two forms of lupus which he terms /

terms lupus minimus, when there is little ulceration, and lupus maximus when the ulcerative process is extensive. He closes his first paper by giving a differential diagnosis between lupus minimus and pruritus Vulvae, caruncle, eczema, and between lupus maximus and cancer, syphilis and elephantiasis.

In his second contribution to the study of esthiomène entitled: "On the Ulceration of Lupus on the Female genital organs including Pits, Perforations and Excavations" (Transactions of the Obstetrical Society 1885) Mathews Duncan relates the histories and describes the conditions of five cases of lupus vulvae. He uses the term lupus in preference to esthiomène "since it expresses the great eroding character of the disease, including ulceration, inflammation and hypertrophy variously combined, not cancerous not epitheliomatous, not syphilitic." Apart from stating again that the condition is not of a scrofulous nature he does not enter into a discussion on the aetiology. From the outset he considered that lupus of the face and lupus of the vulva in spite of their physical resemblance to each other were probably of different origin.

"The /

"The name of lupus is retained in order to avoid change and because the character of the disease brings it into alliance with the ordinary run of such diseases."

An interesting analysis and full report on all cases published under the names of Esthiomène or Lupus Vulvae up to the year 1887 was published by Dr. Grace Peckham Murray of New York. The author contributed herself two monographs to the study of esthiomène. Her first dissertation on the subject appeared under the title: "A contribution to the study of Ulcerative lesions of the Vulva commonly called Lupus or Esthiomène" in the American Journal of Obstetrics in 1887. The author describes in detail a case she had under observation for many years and which comforms to the type described by Huguier as "Esthiomène hypertrophique et végétant." Dr. Grace Peckham Murray then proceeds to classify the cases published as lupus or esthiomène according to their various aetiological factors and the clinical conditions found, giving a complete survey of all that was hitherto written on the subject of Esthiomenic ulcerations from the year 1849 when Huguier published /

published his essay to the year 1887. She found that many constitutional diseases coexisted with esthiomenic ulcers, many being apparently directly causative of the vulvar ulcers; in fully half the cases there was a clear history of syphilis; in many there were indications of tuberculosis and some suffered of cancerous ulcers concurrently.

"A hitherto undescribed form of New Growth of the Vulva" is the name of an essay published by R. W. Taylor M.D. in the "Journal of Cutaneous and Genito Urinary Diseases" in 1889. The author describes two cases of this "new growth" which he considers to be distinct from syphilis or gonorrhoea but which "might be mistaken for lupus". He states that in one case the new growth supervened on "a chancroid lesion" but was quite "benignant in nature", showing only inflammatory tissue.

It is from the histological examination of these two cases which showed purely inflammatory tissue that the author considered himself justified in diagnosing them as cases of a "hitherto undescribed form of new growth". The same author in his text book on "Sexual Disorders of the Male and Female" (1897) /

(1897) gives his views on Vulvar hypertrophy with ulceration and mentions that the following factors are the causes:-

1. Recurrent attacks of Herpes.
2. Constant irritation and inflammation.
3. Trauma.

After describing the various forms of Vulvar hypertrophy R. W. Taylor proceeds to describe the formation of ulcers on the hypertrophied parts. He considers them to be purely accidental and secondary manifestations caused by pressure or trauma on coadaptation of parts. "They have often been described as lupus although they are not of a tuberculous but of a simple nature." He states that these ulcerations tend to keep up the morbid process producing deep fistulae and atresiae and finally causing death. He does not explain how ulcers of a simple and secondary nature can burrow so deeply and cause death by widely destructive changes.

There can be no doubt that R. W. Taylor in describing these hypertrophies with ulceration which he termed in his paper " a hitherto undescribed form of new growth" are esthiomenic in nature.

Numerous /

Numerous contributions to the study of esthiomène have been published by German authors under the name of "Ulcus Vulvae." Virchow was the first to introduce this name and Veit in his "Handbuch der Gynoecologie" 1898 adopted it. He gives a rapid survey of the greater part of the literature published on the subject of vulvar ulcerations with hypertrophy and discusses the various aetiological factors and theories. He considers that in all cases of Ulcus Vulvae, syphilis has preceded, adding that the ulcer itself is however not specific in nature. In describing a case of Ulcus Vulvae, he follows the description given by Schroeder in his essay on "Ulzerazionem an der vorderen und hinteren Kommissur."

The last of the important publications on esthiomène in point of time is S. Pozzi's chapter on esthiomène in his "Traité de Gynécologie clinique et opératoire" 1907. He describes in detail the two forms, mainly hypertrophic and mainly ulcerative and agrees with the earlier French writers Bernutz and Figuet in assigning the condition to tuberculous infiltration. He holds that directly or indirectly esthiomène /

esthiomène is always tuberculous in nature although the Bacillus of Tuberculosis has only once been found (by Martin et Nicolle). He does not enlarge on the histological findings of his own and other people's cases and his conclusions are therefore not convincing.

Some of the most recent studies on the subject under discussion were published by Dufuy et Rullier, (1907) St. Lazare Hospital, Paris who term the condition a simple scleroma with ulceration neither tuberculous nor syphilitic in nature but simply due to local irritation and uncleanliness. Their observations have not been published in any thesis or monograph.

CHAPTER III.

Morbid Anatomy and Histology.

The two main features that all cases of esthiomène present in a varying degree are ulceration and hypertrophy; one or other of these features may be greatly in excess over the other but unless both features are present the case is not one of esthiomène. Hence a large number of cases published as esthiomène which showed only ulcerations must be excluded from the records. Many of these are obvious tuberculous ulcers. The degree of hypertrophy and ulceration depends on the stage of the disease. For instance in the case of Ellen Errington there was a small fleshy body around the urethra which was at first considered to be an angiomatous caruncle and two small inflammatory patches in the vagina on the point of breaking down into ulcers. This constitutes a very early case and corresponds to the first type described by Huguier as "Esthiomène erythémateux et tuberculeux."

At a later stage the ulceration and particularly the hypertrophy are more marked. The whole vulva may be greatly deformed and distorted; any part of it may /

may be affected, but the clitoris and labia minora suffer more frequently than the other structures.

Large firm masses project irregularly from the general surface of the vulva and often greatly outside the labia majora. These masses represent definite vulvar structures which have undergone inflammatory changes. There is usually no sign of the structures themselves nor can the parts be differentiated from each other if several structures are affected at one time. These masses are not unlike the combs and wattles of birds and are so peculiar and distinctive in their appearance that they produce a lasting impression and make it impossible for the observer to confound this form of hypertrophy with other hypertrophies such as elephantiasic or tuberculous which look and feel different. The hypertrophied masses of esthiomène are sometimes pedunculated but more often sessile; they are so irregular that sometimes one part may be sessile and another pedunculated. Their surfaces are usually very uneven, tuberculated, torn and jagged. In the deep fissures between the irregular convolutions a process of ulceration is often going on which may lead to /

to fistulae without showing any surface ulceration at all. In the case of Maggie Lawrence the hypertrophy simulated a bunch of grapes; the mass projected boldly from the subjacent tissues.

The hypertrophied masses are firm and elastic to the touch and quite painless. The skin over the masses is usually dry and hard and is often of an almost dead white colour; as a rule the colour is pinkish yellow contrasting with the purply brown of the other vulvar structures. When the hypertrophy is extensive, pressure sores may form on the surface but these are rare, much rarer than in Elephantiasis Arabum because the hypertrophy never reaches very large dimensions. The integumentary structures around the mass often show purplish or brown colour changes and sometimes are bright red (esthiomène erythémateux.) The ulcerations vary considerably; there is not a definite type with a typical floor and characteristic edges. They may be shallow or deep, sloughy or dry, red purple or brown. They are always irregular in outline, and may be shallow in one part and excavating in another. The edges may be thick or thin, sloping or vertical, undermined or rolled /

rolled, white grey or red; in fact every type of margin has been observed and reported. When the ulcers are shallow they are more likely to be extensive, when they are deep they have small, sometimes hardly visible orifices on the surface. In the case of Patience Pearcy the orifice of a deep ulcer was covered by a flap-like valve.

It appears that in such cases where the hypertrophy is very marked the ulceration is not as a rule very extensive. In the case of M. Lawrence it was very slight; there were only two areas of shallow ulcerations.

The third stage may be said to be the stage of actual destruction. This destruction is due to deeply burrowing ulcerations causing fistulae between the body cavity the visceral organs and the external surface. These fistulous tracts may heal in part and the result is stricture formation producing atresiae of vagina urethra and rectum. In some cases reported by the French there was enormous destruction of all the tissues.

It is remarkable that in this severe form of the disease the ulceration process is greatly in excess /

excess over the hypertrophic one; in fact in some cases like, for example, Patience Pearcy the hypertrophy is limited to one small area and may consist only of a "firm and projecting area growing from the lower part of the Vagina." In her case both fistulae and strictures were present.

The cicatrices produced are very firm unyielding and large in amount. Their contraction produces very marked deformity. When they appear on the surface they are tense shiny with an even surface: their chief characteristic is that they are very apt to ulcerate again after a time so that the process of disease may go on for many years.

Histological Findings.

A comparatively small number of the cases published as esthiomène have been examined microscopically. This is no doubt due to the fact that in the days when esthiomène was a subject of frequent controversies the histological methods were troublesome and inefficient. In most cases where the tissues were examined microscopically, connective tissue alone was found, without new growth and without pathogenic organisms where such were stained for. This led to the view on the one hand that the condition called esthiomène was a purely inflammatory one due to irritation or trauma and on the other hand that it was a tertiary syphilitic manifestation.

The first pathological report on esthiomène in the annals of literature is the one of M. Robin who examined 4 cases of Huguier and attached them to Huguier's essay of 1849. This is what he says: "Un epithélium normal, le tissue subepithétial est épaissi d'un centimêtre, élastique blanchâtre et crie sous le scalpel. Les tissues cellulaires n'offrent rien de caractéristique. Ce sont des tumeurs /

tumeurs homéomorphes, c'est à dire composées par des éléments anatomiques qui se retrouvent dans les tissus normaux de l'économie."

Cornil made microscopical examinations of the specimens removed by Bernutz from his cases and he found marked alterations of lymphatics and hypertrophy of the papillae. The blood vessels of the papillae were engorged. In the deeper parts of the subcutaneous tissues he found a large amount of new connective tissue with long fine spindle shaped connective tissue cells of a new "embryonic nature." The lymphatic cells were dilated and their walls considerably thickened.

In 1876 Figuet published his "thèse de Paris" on esthiomène and appended to it the report of Troisier on the microscopic findings. He found the epidermis for the greater part normal. The walls of the vessels in his subcutaneous tissues were enormously hypertrophied dilated and engorged: they showed numerous embryonic connective tissue cells in their vicinity. The deeper tissues showed quantities of new connective tissue, with dilated vessels which were surrounded by large masses of round /

round cells.

Mathews Duncan published two series of cases of esthiomène; the first series was published in connection with his first paper on the subject (On Lupus of the Pudendum 1884). The second series appeared in 1885 with his second paper("On the Ulceration of Lupus on the Female Genital Organs including Perforations, Pits and Excavations").

Dr Thin appended his microscopical report to both papers. In the first paper he states that no new tissue elements were found but young growing fibrous tissue with many leucocytes grouped round the vessels. He finds that the tissues differ from lupus in the absence of tubercle foci. Dr Thin in examining M. Duncan's second series of cases found "the usual characteristics of an elephantiasic hypertrophy". They were all cases of a "low chronic form of inflammation."

Deschamps (1885) found the same histological features in the esthiomenic tissues he examined as Troisier. "Une espèce d'infiltration hypertrophique de la peau, particulièrement du derme, altération des vaisseaux lymphatiques; un épaississement du corps muqueux /

muqueux de Malphigi, des colonnes descendantes inter-papillaires. Les papilles hypertrophiées vaisseaux papillaires enlargis et le tissu de la papille enfiltré d'éléments embryonnaires et de leucocytes. Enfin le tissu sous-dermique qui comprenait toute l'épaisseur de la tumeur contenait une quantité de tissu lâche, vaisseaux dilatés entourés d'une quantité d'éléments embryonniques."

R. W. Taylor M.D., N. York the author of the essay on "A hitherto undescribed form of New growth of the Vulva" in the Journal of Cutaneous and Genito-Urinary diseases 1889 described the morphology of his New Growths which were undoubtedly esthiomenic hypertrophies with ulcerations "as simple inflammatory tissue."

The two valuable essays written by Dr Grace Peckham Murray of New York are both accompanied by microscopical reports on the cases published. The first is entitled: "A Contribution to the study of Ulcerative Lesions of the Vulva, commonly called Lupus or Esthiomène." It appeared in the American Journal of Obstetrics in 1887. The microscopic report by Dr Coe is appended. He found the epidermal structures /

structures absolutely normal but an extensive round celled infiltration was seen in the corium, "the cells being similar to those seen in ordinary granulation tissue. They are arranged in groups which occupy the interstices of the connective tissue and are frequently seen surrounding dilated blood vessels." There is general dilatation of the lymphatics. In some parts the cell infiltration is very marked. There are no giant and epithetioid cells. "In short the microscopical appearances are those of simple inflammation of connective tissue with this peculiarity that the round cells show a decided tendency to form circumscribed groups or nodules."

At a later date Dr. Grace Peckham Murray published her "Second Contribution to the Study of Ulcerative lesions of the Vulva commonly called Lupus or Esthiomène" to which Dr. Louise Cordes added her bacteriological report. She found a streptothrix which was pathogenic to rabbits and white mice and was obtained in pure cultures from the surface of the ulcer. The epithetium in the neighbourhood of the ulcer showed mycelial threads of this fungus. The bacteriologist did not draw any conclusions as to the importance /

importance of this streptothrix as an aetiological factor. (American Journal of Obstetrics Vol. 47.)

In 1903 André Boursier in his book on gynaecology entitled: "Précis de Gynécologie" and again in his essay in 1908 "On hypertrophic ulcerations of the Vulva" which appeared in the Journal de Médecine de Bordeaux reaffirmed the purely inflammatory nature of esthiomène and reported his histological findings as follows: "L'épiderme épaissi envoie dans le derme des prolongements épidermiques interpapillaires. La masse de la tumeur est faite par un tissu conjonctif à fibres ondulés contenant une grande quantité de cellules en amaz surtout autour des vaisseaux. Ceux-ci, surtout les lymphatiques sont tres dilatés et sont gorgés de celles blanches et rouges. Les vaisseaux sont quelquefois si dilatés que la tumeur est caverneuse. Dans les tissus sous-dermiques on trouve aussi des vaisseaux dilatés. Il n'y a que peu de tissu élastique."

It is interesting to note that those cases where tubercle foci and bacilli were demonstrated (Martin et Nicolle, Günther and others) although published as esthiomène did not present the characteristics necessary /

necessary to constitute cases of esthiomène. In more recent days when the demonstration of the Bacillus of Koch is easily carried out the cases were usually published as "tuberculous affections of the Vulva." Some of the older writers have seen giant cells and epithelial cells and upon those findings they based the assumption that esthiomène is the same as lupus of the face.

We know now that epithelioid and giant cells are not pathognomonic and we therefore do not accept the conclusion of these authors that their cases of esthiomène were of a tuberculous nature as correct. A few cases have been diagnosed as cancer or rodent ulcer. It is possible that these cases like the case appended to this thesis (Maggie Lawrence) showed cancerous degeneration of a tissue previously hypertrophied and ulcerating or that they were from the outset epitheliomatous or sarcomatous.

The view that esthiomène was a malignant degeneration attended by hypertrophy and ulceration was based on theory rather than on fact, for the histological reports of those who maintained the view were not published. All the evidence we have goes to show that /

that esthiomène is not a malignant disease in the usually understood sense of the term (i.e. there are no new tissue elements) and that it is not lupus in the modern meaning of the term, but that all the tissues are purely of an inflammatory nature.

The histological examination of the tissues of 4 cases appended to this thesis confirms the view that esthiomène is an inflammatory condition. All the features found by those who studied the morphology of esthiomène are typically represented in the case of Ellen Hill. Inflammatory tissue of old and of recent date is found showing all the stages from an early acute stage to a late one of dense fibrous tissue formation. The state of the blood-vessels and lymphatics of the dermal papillae and surface epithelium point to the long duration of the inflammatory process. The numerous dense clusters of round cells found chiefly around the blood-vessels indicate that the inflammation is still proceeding.

In the slides appertaining to the case of Ellen Errington the inflammatory tissue is of recent date. The case was a very early one. In the case of Maggie Lawrence we found among the greatly increased /

increased fibrous tissue groups of malignant cells. These are limited in number and space showing that the malignant degeneration had supervened on the inflammatory one at a very recent date.

Maggie Lawrence had suffered for some time (3years) from "a breaking out on the Vulva" namely ulceration with hypertrophy. On admission there was a large hypertrophied mass, two shallow ulcerations and a small nodule in the skin in the vicinity of the hypertrophied mass. No features indicative of malignancy were present as the ulceration was very superficial and did not show the usual characteristics of a malignant ulcer. Typical cancer cells were found under the microscope. We conclude that malignancy supervened on esthiomène.

There can be no doubt that the reputation of great fatality esthiomène had among the older writers was partly due to the fact that malignancy occasionally supervened on a case of some standing. There are however no microscopical records that such was the case. Brodie, Cooper and Paget held that the condition was a malignant one and so did Macnaughton Jones /

Jones who stated that "lupus may be considered an epithelioma."

Bacteriological Examination.

Amongst the older writers there is only one, Dr. Louise Cordes who made a close and detailed bacteriological examination. As we have already seen she discovered a streptothrix on the surface of the ulcer with projecting mycelial threads among the epithetial cells. She did not draw any conclusions as to the causal agency of the organism. It was probably merely an organism accidentally present on the floor of the ulcer.

Before the discovery of the Bacillus of Koch no reports on bacteriological examinations of esthiomenic tissues were published but after that event a number of sections with the tubercle bacillus insitu were published especially by those who maintained that esthiomene or lupus vulvae and lupus elsewhere are the same disease. No publication regarding the presence of the Spirochaeta Pallida in esthiomenic tissues has yet been made.

In the case of Ellen Hill a detailed bacteriological examination has been made. Tubercle Bacilli were stained for by the Ziebl-Nielsen method of staining and other micro-organisms with the ordinary acid and alkaline dyes. No micro-organisms were found.

CHAPTER IV.

Clinical Features of Esthiomène.

Type of Subject:

Esthiomène is most frequently found in women between 20 and 40 though occasionally it is met with in the elderly. It occurs more often in the unmarried than in the married. Very rarely are the patients mothers of large families. They usually have had miscarriages, premature births and sometimes one or two children who died in infancy. When the disease occurs late in life we find more frequently that the patient has had a number of healthy children and no miscarriages. Esthiomène does not often occur under 20 and only occasionally after 55.

Laundresses and domestic servants furnish a good number of cases of esthiomène. They are often of the poorest and most neglected section of the population and many earn their livelihood in a precarious way. This is not however constantly the case for, although all cases of esthiomène reported are from the "hospital class of patient", many are well cared for and are cleanly in their habits and regular in their mode of living.

In /

In Germany and France a greater number of the severe cases of esthiomène have been seen and reported and in both countries the authors on the subject lay great stress on prostitution and a life of misery and neglect. We have no doubt that there is a direct relationship between that and the severe cases of esthiomène. This is discussed in the chapter on aetiology.

Condition of General Health.

Subjects of esthiomène are often described as blooming robust young persons and statistics appear to show that on the whole the general health is not much affected. Many authors have expressed surprise that with so much destruction often with strictures and fistulae there should be so little constitutional disturbance. Dr. Grace Peckham Murray's patient though suffering from deeply spreading ulcers went about with heavy baskets all day plying her trade as a pedlar. Mathew Duncan especially points out that the women are often extremely healthy in appearance. Of the 33 cases collected by Dr. Grace Peckham Murray 21 enjoyed good health, 5 had poor health and 7 were not reported. The state of health depends of course on the stage at which the disease has arrived. When there is a fistulous tract into the rectum with constant escape of this faeces or into the urethra, the health degenerates rapidly. Intercurrent attacks of slight pelvic peritonitis occur later which make the patient a complete invalid.

Symptoms.

The insidious nature of esthiomène is well known. For many months hypertrophy and ulceration may be present without the patient being aware of the condition. There is a characteristic absence of burning darting and pricking sensations in the early beginnings. The patient may have known for some time that there was a "lump" present in some part or other of the vulva but she has not taken any notice until some day a discharge appeared which made her seek medical advice. Often advice is sought only when the ulceration is getting very extensive or when itching becomes troublesome. Comparatively few cases are seen at a very early stage so that the actual beginnings of the condition are not known. An exception to this is the case of Ellen Errington (refer) who had only two small inflammatory patches and a hypertrophied lump of tissue round the urethra. Even when the disease has well advanced there are few local disturbances; there is incontinence of faeces and urine owing to fistulous burrowings of the ulceration but very little pain or none. It has been noted that the discharge from esthiomenic ulcers is /

is scanty in amount and has either no odour or a very slight one which is not offensive.

Local Conditions.

We have already discussed the various appearances presented by cases of esthiomène. Many varieties and subvarieties have been described, the best descriptions and classifications being those of Huguier. There is not one special and definite type; many appearances noted by some authors are not seen by others. The variety depends on the stage of the disease, as we have already pointed out. For instance there is no "perforating type" which perforates from the outset of the disease, but most cases finally arrive at the perforating stage if the case is neglected or does not respond to treatment. Like Huguier we distinguish between the purely superficial inflammatory and sometimes tuberculated type, the destructive type which tends to burrow in the subcutaneous tissues and to undermine the healthy as well as the hypertrophied areas, and finally the perforating type which produces fistulae and strictures. The three types are synonymous with the three stages observed in the course of the disease; they gradually pass from one to the other and show no distinctive intervals.

History of Previous Health.

The disease which is most frequently found in the previous history of patients suffering from esthiomène is syphilis. The number of cases of syphilitic infection (usually some years previous to the first appearance of esthiomène) is very striking. Many patients have a distinct history, others show indications of their having had syphilis. The next disease in frequency is tuberculosis. Sometimes there is phthisis present and syphilis as well; sometimes there is lupus of the face or other parts. Ulcerations of the vulva in such cases are so different from esthiomène that they can easily be distinguished from that disease. Trauma and abscesses are sometimes reported but by far the most frequent cause of previous ill health is syphilis.

Enlargement of glands in the groin.

In a large percentage of cases of esthiomène where a clear history of syphilis is obtained the glands in the groin are reported as enlarged. An absence of enlargement however does not exclude syphilis. They may show no enlargement and yet syphilis may have been present. When they are enlarged they support the diagnosis.

Duration, Course and Prognosis.

One of the distinctive features of esthiomène is the prolonged course it runs. This is due to the following facts:-

(1) The general health is not seriously affected until the pelvic viscera show signs of ulceration. This occurs late in the history.

(2) The disease is extremely chronic in its nature; acute ulcerations are not met with. In this respect esthiomène resembles the callons chronic ulcer of the leg which may persist throughout a lifetime.

(3) The tendency towards healing and citrasition; one may see a large mass of cicatricial tissue in one part while in another there is a new ulceration in progress. This healing is however not permanent for the cicatricial tissue is very apt to break down again and to become the seat of a new ulceration. In this way the condition may go on for an indefinite time.

(4) Occasional temporary response to rest and treatment with subsequent relapses.

C. N. one of the cases attached to this work suffered /

suffered from esthiomène for 11 years; another case, reported by Cayla was seen when it had lasted 11 years; two others of 8 years standing are reported by Angus Macdonald and Peckham Murray. Cases of long duration are usually reported to have ended in death. One death occurred 15 months after the beginning of the illness which would appear to be exceptional. The intractable cases go on for a very long time and finally end in death from exhaustion. Those that respond to treatment usually do so in the first or second year after the onset. Many are considered cured where a small area of ulceration may have persisted and the condition may begin de nuovo after a time. Nevertheless in a majority of cases the recovery is permanent and complete. When the condition has gone on for some years and does not show response to treatment nor real attempts at cicatrisation and temporary improvements the outlook becomes grave. When excavations and fistulae have actually begun there is no likelihood of recovery. Such cases end in death after a long time of exhausting diarrhoea. Intermittent attacks of pelvic peritonitis accompanied by attacks of vomiting and diarrhoea with febrile temperature /

temperature occur. These cases however are getting rarer and in these days when the nature of the disease is beginning to be recognised they are hardly ever met with. A typical case is C. N. The greatest number of cases yield to treatment in the earlier stages of the disease and a good number become cured at a later stage. There is a danger however that in the course of the illness the tissues may undergo malignant degeneration. This has not been noted by those who reported their cases. No doubt it is a comparatively rare occurrence but esthiomène is altogether a rare condition. It is likely that those who found a hypertrophied state of the vulva with malignant ulceration put it down as carcinoma from the outset without taking note of the fact that the hypertrophy with ulceration may have existed for some time and that the malignant degeneration may have been a matter of recent occurrence.

Epithelioma and squamous carcinoma of the vulva present totally different characters from esthiomène and there should be no difficulty in distinguishing the two from each other or the former from a malignant degeneration of esthiomenic tissues.

CHAPTER V.

I. 1908.

Case of Maggie Lawrence.

Age 35. Married.

Occupation: Pressworker.

Diagnosis on Admission: Esthiomène.

Personal History:

Patient was married at the age of 18.

A miscarriage at the second month took place in the first year of her married life.

Soon afterwards she had another miscarriage at the sixth week.

In the following year she was delivered of a full term child which was perfectly healthy until its eighth month when it died of bronchitis.

Since then the patient has had no children and no miscarriages.

History of Previous Health:

Patient was perfectly well until she was 15. She then began to work in an enamel factory and from that time onwards had frequent coughs and colds in the head.

After her marriage at the age of 18 she began to /

to feel ill and she has never been quite well and strong since then. She has been troubled with frequent coughs and colds and was told "that her heart and lungs were weak."

At the age of 22 had a severe attack "of pleurisy and inflammation of the lungs;" she returned to work five weeks afterwards. Patient states that she has been short of breath ever since.

Seven years ago began to suffer severe and constant pain in the back. It sometimes shot through to the iliac region but as a rule it was limited to the back. Patient sought advice at the Queen's Hospital and saw Dr. B. His treatment did not improve her condition and she went some time later to the Orthopoedic Hospital where she was put into a Plaster of Paris jacket which she wore for two years and which greatly relieved her pain. A year ago she came to the General Hospital for treatment for her cough; this improved after treatment. Her back still pains her a good deal.

Menstrual History:

Began to menstruate at the age of 15. With one intermission of nine months when she was feeling ill, she /

she has always been regular. Type: 21 days. There is some discomfort but no pain.

History of Present Complaint:

Patient consulted Dr Thomas Wilson on account of itching and soreness of the vulva. The itching is worse at night. There has never been any similar trouble before.

Pain in the vulva and surrounding parts comes on chiefly after walking; as a rule it is localised but sometimes shoots into the right groin.

Both pain and itching are greatly increased during menstruation and micturition. No indication of urinary difficulties. There is some constipation. Patient has noticed an occasional slimy bloodstained discharge from the vulva; this discharge only comes on on walking and standing; the parts are more painful when they are discharging. The discharge is odourless and small in amount. Menstruation regular.

General Condition.

Patient is thin and spare and looks delicate; marked malar flush, some cyanosis of lips and buccal mucous membrane. Finger tips, especially those of the right hand are clubbed.

Patient /

Patient does not suffer much pain or inconvenience at the present time.

Examination. 24th Oct. 1908.

Lungs and heart appear perfectly healthy.

Back: No curvature.

On making deep pressure of the laminae on the left side in the interscapular region tenderness is elicited. The vertebrae affected are the 3rd 4th 5th and 6th.

Inspection of the Vulva:

A mass of hypertrophied tissue is visible in the right half of the vulva. It represents the clitoris the hood of the clitoris and the right labium minus. Not one of these structures can be distinguished from the mass. The tissue is firm elastic dry and fleshy to the touch. It stands out in bold relief from the centre of the vulvar structures. Its surface is convoluted rough wavy, resembling the combs and wattles of birds. The colour is yellowish pink, white in parts. The mass is sessile. The surface tissue is not ulcerated. The other parts of the vulva are of normal size and contour.

Two areas of ulceration are visible; one area

½ /

½ x ¾ in. in size is situated at the anterior end of the left labium majus; the ulcerated area is red and raw, discharging slightly. It is quite superficial. Its margins are only faintly marked.

The other ulcer is placed to the outer side of the hypertrophied area near the posterior end of the fleshy mass. It is quite superficial and presents the same features as the other ulceration. (See accompanying colour sketch from life by the author).
Vaginal examination: Nothing abnormal to note.
Inguinal regions: No scars; one enlarged gland.
Operation: Excision of all the ulcerated and hypertrophied parts of the vulva. Removal of inguinal glands on the right side.
Specimens:

(1) One specimen consists of the right labium minus the preface and clitoris, the vestibule.
(2) One Specimen consists of the right inguinal glands.

Specimen 1.

There is a margin of a healthy skin area ½ in. wide around the hypertrophied parts.

The colour of the mass is of a dull opaque white; its /

its surface is thrown into blunt papillae. On these small papillae are points of red colour, where the surface epithelium appears to be just giving way.

The ulcerated masses show unhealthy granulations. A small nodule is visible in the neighbouring skin around the anterior pole of the hypertrophied mass, distinct from the latter.

This nodule, part of the hypertrophied area and the enlarged gland were used for microscopic sections.

Microscopic Examination.

The section of hypertrophied part, nodule and gland show typical squamous celled cancer.

The stratum corneum is thinned and has disappeared in parts; large masses of darkly staining proliferating epithelium grow in solid pillars into the subjacent tissues; numerous epithelial cones and typical keratinous whorls are seen lying detached or in groups in the subcutaneous tissues.

The gland shows cancerous infiltration.

The accompanying colour sketch shows the condition of the vulva; the microphotographs represent sections of the nodule showing cell nests.

Observations.

1. History with regard to reproductive system points to syphilis or gonorrhoea, namely two miscarriages and "one child sterility." There is however no history of any luetic or gonorrhoeal infection.
2. History with regard to respiratory system and spine point to possibility of tuberculous infection. The patient looked tuberculous but the lungs were found healthy.
3. The hypertrophy and ulceration of the vulva is typical of the disease described as esthiomène. The question arose whether the cause of the condition was syphilis or tuberculosis or whether a merely accidental inflammatory process had taken place. The microscope revealed typical squamous celled carcinoma. It is most unusual for that disease to cause a marked hypertrophy with shallow ulcerations in the non-hypertrophied parts. It is possible therefore that the condition was primarily due to syphilis and that cancerous degeneration occurred in the hypertrophied areas.

Colour Sketch.

Copied from life by the author.

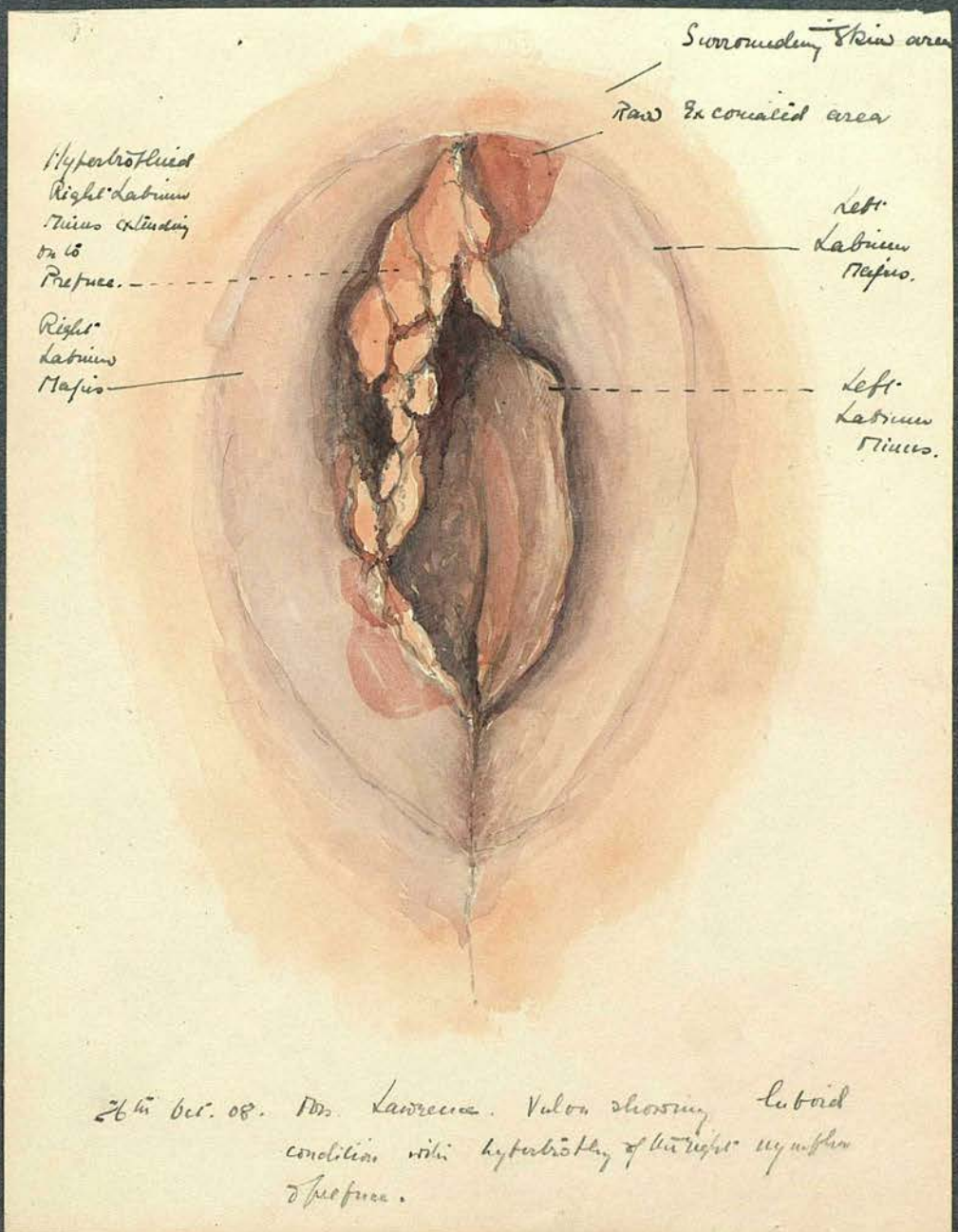
Maggie Lawrence.

26th October 1908.

Vulva showing "lupoid" condition with hypertrophy of the right nympha and prepuce.

The other parts are not altered in shape and condition but presented a perfectly normal appearance.

The two ulcerations, one to the left of the clitoris and the other to the right and in the front of the perineal body were shallow and discharged a thin watery clear fluid.



26th Oct. 08. Mrs Lawrence. Vulva showing clitoroid condition with hypertrophy of Right Nymphae & Prepuce.

Maggie Lawrence
Colour sketch from Life.

Section I.

Maggie Lawrence.

(Low Power.)

The section shows typical epitheliomatous tissue. The epithelial cells grow downwards into the subcutaneous tissues in the form of solid phalanx. The usual relationship between the skin layers is lost.

There is comparativly little small-cell infiltration.

In the midst of the obliquely cut column of the epithelial cells seen in this section there is a 'pearl' or whorl of compressed corneal cells.

The cells in the immediate vicinity of this pearl show some disintegration.

Maggie Lawrence
Section I. Low Power.

Section II.

Maggie Lawrence.

(High Power.)

The same section magnified shows the column of cancerous cells and in their midst the corneal whorls from which arises the German term "Hornkrebs."

Maggie Lawrence. Sec. II. High Power.

II. 1898.

Case of Lucy Blair.

Age 28. Married.

Personal History:

Married 8 years ago. One child born three months after marriage; lived two days; no miscarriages; no pregnancy since. Labour at term, normal.

History of Present Complaint:

Patient complains of something in the seat which causes much pain on sitting down and discomfort on standing and walking.

History of Present Condition:

Two years ago noticed a swelling near the anus. This swelling has been increasing gradually; six to seven months ago it began to discharge; the discharge was brownish, sometimes blood-stained and had a bad odour. The pain came on gradually.

History of other illnesses:

She has had "bad sore throats several times. She suffers from severe headaches and has a rash on the arms from time to time." Has had no other illnesses.

Catamenia:

Catamenia:

Began at sixteen. Has always been regular.

Family History:

Father and mother alive and well. Four brothers and one sister ditto.

Husband has been a brass-caster and hawker alternately since he was fifteen. Five years ago he was in the Army for 1½ years but was discharged.

Patient was first seen on September 1898.

The condition was then diagnosed as a Condylomatous Hyperplasia of the anal and vulvar tissues.

In February 1899 she was admitted into Ward 15 of the General Hospital.

Condition on admission:

A nodular hard swelling with a scar is present in the right groin; enlarged glands in the left groin.

Vulva: At the left margin of the anus, there is a nodule about the size of a hazelnut, firm and inelastic. The skin around the anus is thickened, red and moist. There is a small ulceration on the left side of the anus, a depression and fold of skin on the right. At the posterior part on the inside of the right labium majus is an irregular ulcerated area /

Treatment.

Inunctions of Unguentum Hydrarg. ℥ss given daily until salivation occurred. Then she was put on Potassium Iodide.

Parts around the vagina and anus were quite healed at the end of February 1899. It was difficult to introduce the finger into the anus on account of thickening and hypertrophy of sphincter.

A small amount of tissue had been removed from the ulcerated and hypertrophied areas, for examination.

Observations.

1. A very definite history of syphilis, manifesting itself by sore throats, rashes, headaches, "one child sterility."
2. A combination of marked hypertrophy with ulceration, typical of the condition described as esthiomène.
3. It is interesting to note the rapid response to antisyphilitic treatment: syphilis had been contracted 8 years before treatment, but the symptoms were those usually seen in the secondary stage. The case was fairly acute; healing took place at once. A light stricture however remained.

Microscopic Examination

The parts removed showed the usual dense inflammatory tissue associated with tertiary syphilitic lesions. The dermis and subjacent tissues are infiltrated by large masses of flasma cells; the stronea is dense, the vessels are markedly thickened. The various layers of the true skin have lost their usual inter-relationship, the corneal layer being thinned and the stratum malpighii, showing enormous increase. Solid pillars of slightly oedematous prickle cells are seen to extend into the tissues below. There is however no sign of any tendency towards malignant degeneration.

Lucy Blair.

(Low Power)

This microphotograph shows the usual features of syphilitic inflammatory tissue.

Density is its main characteristic; even at a comparatively early stage when small cell infiltration and vascular engorgement are at their height, there is already marked fibrosis showing itself in the greatly thickened capillary walls and the solidity of the tissue stroma.. This section shows no ulceration. There is no loss of continuity of the surface epithelium. But the epithelium is thickened, oedematous and unhealthy.

The section formed part of one of the hypertrophied areas.

Lucy Blair.
Low Power.

III. 1902.

Case of Ellen Jones Hill.

Age 22. Single.

Was seen for the first time on the 2nd December 1902.

Diagnosis. The condition was then diagnosed as a tubercular or syphilitic ulceration of the vulva.

The patient stated that she had had a breaking out on the vulva for three years.

Catamenia: began at 16, regular, duration 6 days, intervals 6 weeks; no pain, there had been some discharge.

Condition on Examination.

On the inner surface of the right labium majus is a large ulcer with sharply cut edges, 2 inches long, by 2 inches broad. In the base is a thick adherent greyish slough.

Was admitted to Ward 15 in December 1902. She was put on antisyphilitic remedies which brought about some improvement. A small portion of the edge of the ulcer was removed for examination. In May the ulceration was healed in front and later was healing also posteriorly.

Microscopical Examination:

The epidermis is greatly thinned; in some places it has disappeared altogether. A few thin detached lamellae of a fibrous deposit are visible on the surface.

The papillae of the dermis have disappeared, their place is taken by typical, fairly recent inflammatory tissue with numerous vessels with large lamina.

There is some extra vasation of blood. The vessels are thickened. The cellular infiltration is more marked in the deeper structures where the inflammatory process appears to be more marked.

Observations.

1. Absence of history of syphilis tuberculosis trauma irritation and inflammation.
2. The condition improved somewhat under anti-syphilitic treatment. This is as we shall see a common occurrence. As a rule the more marked the ulceration and hypertrophy are, the less do they yield to antisyphilitic remedies.

   In early and slight cases that form of treatment is often found to result in permanent cure.
3. Inflammatory tissues with much atheroma such as found in this case are usually found in syphilitic lesions.

Section I.

Ellen Hill.

Floor of Ulcer. (Low Power)

The whole thickness of the skin both epidermis and dermis is destroyed. There is no surface epithelium left. All the layers of skin and their inter-papillary columns of epithelial cells are replaced by a dense mass of darkly staining small cells lying in a mass of fibrinous material. Immediately beneath this surface layer there is a stratum of young connective tissue with numerous new blood vessels and early spindle cells. The small-cell-infiltration becomes less dense towards the deeper strata which are composed of fibrous tissue with thickened vessels and wavy translucent bundles of connective fibres with occasional nuclei. On the surface the tissue is actively inflammatory; it becomes less and less so as we approach the deeper layers. Nowhere is there a vestige of the original skin elements. Blood vessels are very numerous throughout and show great fibrous thickening in the deeper layers.

Ellen Hill.
Section I. Low Power.

Section II.

Ellen Hill. (Low Power)

Portion of tissue from vicinity of ulcer.

The epidermal epithelium is comparatively thin but shows no disintegration apart from a tendency on the part of stratum corneum to shed.

In the subcutaneous tissues we find occasional outposts of inflammatory foci consisting of a circumscribed mass of young round cells. Here and there we come upon a greatly thickened vessel.

Ellen Hill
Section II. Low Power.

Section III.

Ellen Hill.

Portion of tissue taken from the edge of the ulcer.

(High Power)

The epidermis shows oedema and general disintegration of all its layers.

The corneal layer is thinned and vacuolated.

The cells of the stratum mucosum stain feebly in parts and show broken up nuclei. The normal relationship of the epithelial cells of the stratum mucosum to the corneum is lost.

The dermal papillae are irregular and the interpapillary epidermal columns have varying shapes and grow in different directions. Many of them enclose spaces filled with dermal tissue. There are also groups of large fat globules situated in the midst of the skin layers.

In the subcutaneous tissues we see occasional groups of inflammatory round cells.

Ellen Hill
Section III. High Power.

Section IV.

Ellen Hill.

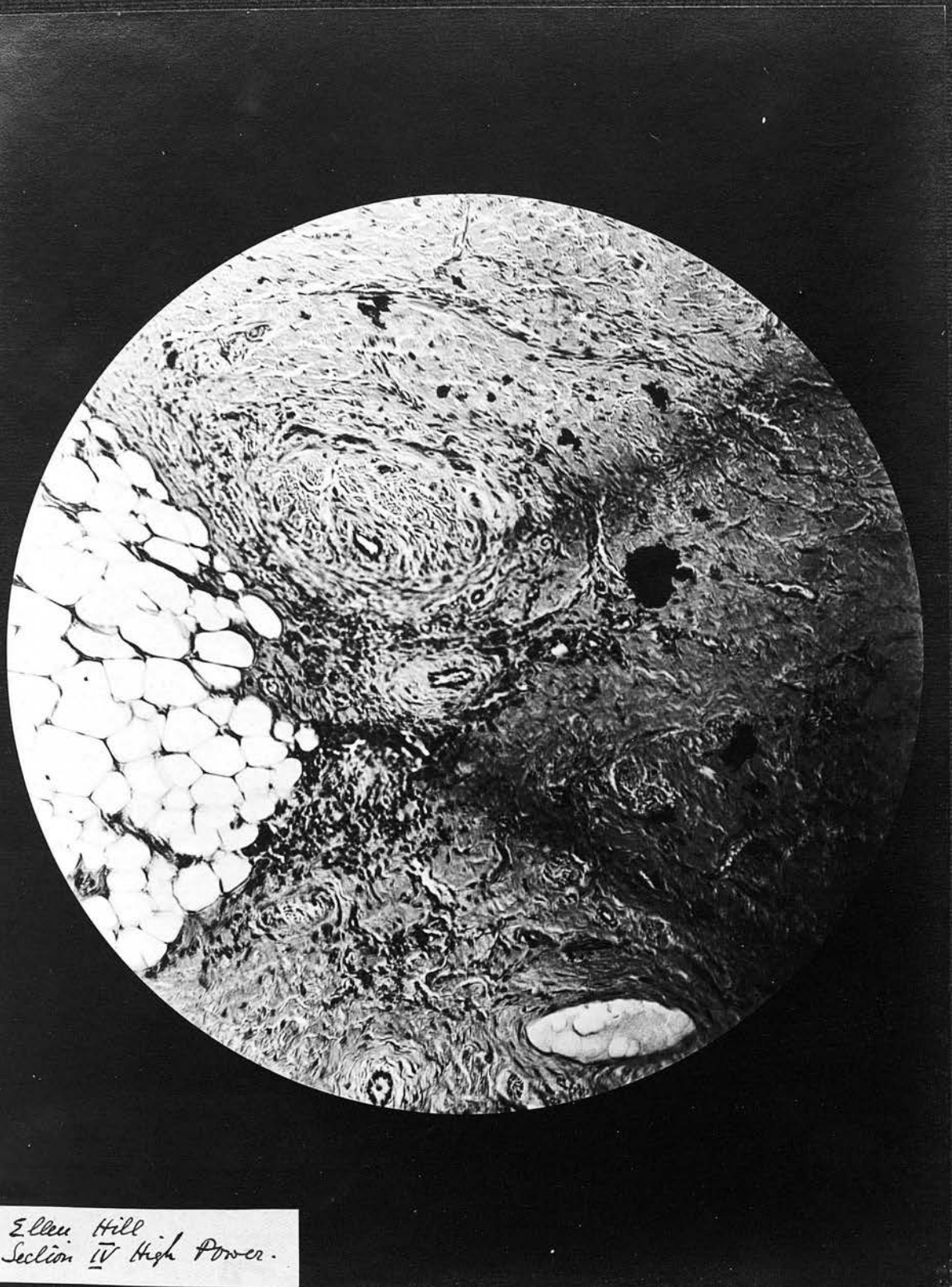
Section from hypertrophied mass of tissues.

(~~Low~~ High Power)

This section shows very dense fibrous tissue of old standing with wavy strands of connective tissue fibres and few cell elements.

There is a small area showing plasma cells. The blood-vessels of the section are exceedingly sclerosed all coats being thickened. The connective tissue is especially dense around the blood-vessels showing a concentric arrangememt of fibres.

Extensive fatty degeneration of the tissues is taking place.



Ellen Hill
Section IV High Power.

Section V.

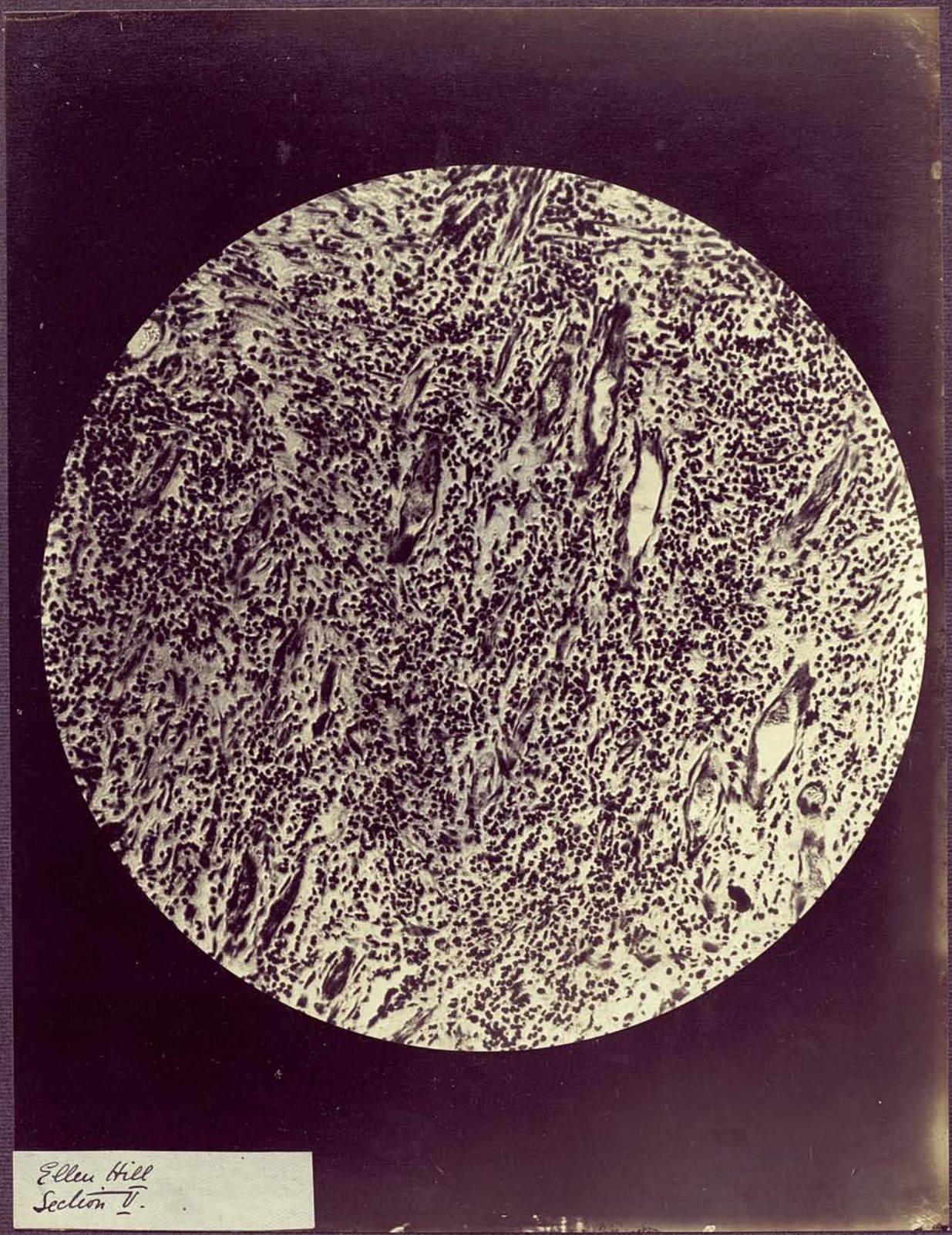
Ellen Hill.

Section from the edge of the hypertrophied mass.

(High Power)

This section shows inflammatory tissue of a later state going on to a further stage of organisation and fibrosis.

The chief elements are the large numbers of small round cells which form very dense masses in some parts, and the numerous blood-vessels. Some of these are widely dilated and engorged with red blood cells: others have thickened walls are surrounded by young connective tissue.



Ellen Hill
Section V.

Section VI.

Ellen Hill.

Blood-vessels showing marked sclerosis and Endarteritis.

(High Power)

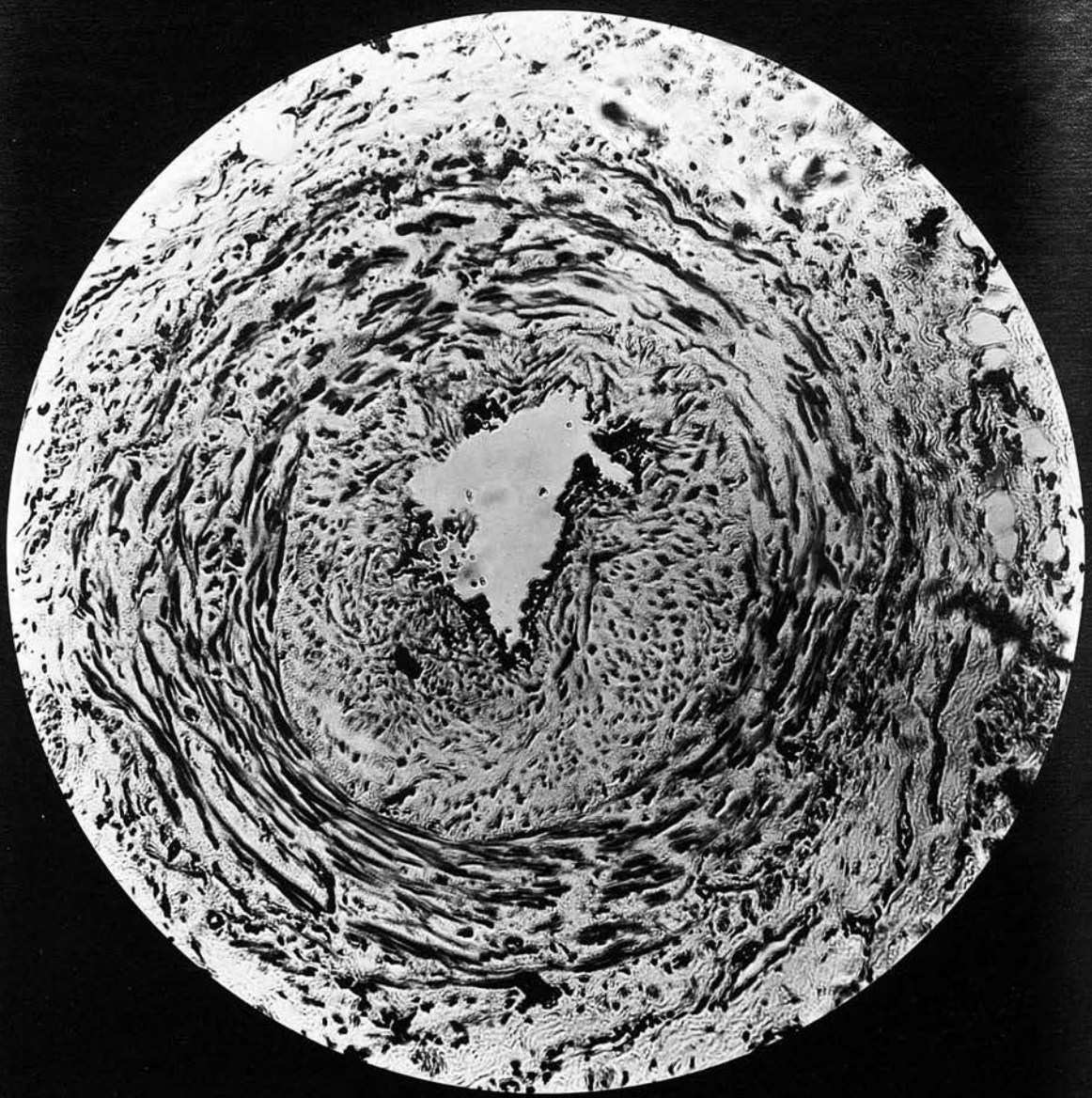
Situated in the deeper strata of the fibrous mass the tunica intima is enormously thickened and tends to obliterate the lumen of the vessel; and the endothelium is disintegrated in parts, showing irregular thickenings which project into the lumen of the vessel.

In some places are completely denuded of endothelial lining. Nowhere do we see normal endothelial cells with clear protoplasm.

Both the middle and outer coats of the vessel are enormously hypertrophied. There is a marked increase of muscular tissue in the middle coat especially marked in the longitudinal layer. The main increase is however due to well organised fibrous tissue whose wall-staining muclei lie everywhere interspersed between the muscular fibres.

The outer coat merges insensibly into the surrounding undifferentiated fibrous tissue and into the fibrosed middle coat from which it is not easily distinguished /

distinguished. A few groups of small inflammatory tissue can be seen lying in the vicinity of the vessel. The general impression given by the vessel and its surrounding tissues is that the process of fibrous degeneration is one of very old standing.



Ellen Hill
Section #1

IV.

C.N. Married, nullipara. Age 24; the wife of a discharged soldier was admitted to the Wolverhampton General Hospital under Dr Thomas Wilson's care on May 31st 1890 with ulceration and hypertrophy of the vulvar and the neighbouring tissues.

Condition on Admission:

She was an emaciated anxious looking woman of medium height and build.

State of Vulva:

The labia majora were greatly enlarged by a solid oedematous swelling and the clitoris was elongated to 2½ in. and was as thick as the forefinger. There was an irregular deep callous ulcer in the vestibule and adjacent anterior vaginal wall; another at the posterior commissure. There was narrowing of the vaginal orifice by firm unyielding tissue; there was stricture of the rectum 1½ in. above the anus, caused by a similar infiltration of its walls and surrounding tissues. Round the anus were several polypoidal outgrowths about the size of marbles. There was incontinence of urine and sometimes of faeces. On the day before admission an attack of abdominal pain with vomiting /

vomiting and diarrhoea commenced and the diarrhoea continued for several weeks. During the stay in Hospital there were several other attacks of severe abdominal pain with vomiting and diarrhoea accompanied by tenderness and by enlargement of the uterus, which could sometimes be felt as large as half a cricket ball above the pubis.

Treatment

Various kinds of treatment for the vulvar condition were tried without effect. Large doses of Pot. Iod. were given, also small and gradually increasing doses, mercurial inunctions and local applications of black lotion.

On Aug. 23rd 1890 patient was anaesthetised with ether; the clitoris and the hypertrophic outgrowths were cut away with scissors and the raw surface and ulcerations burnt by Paquelin's Cautery; at the same time the rectal and vaginal strictures were dilated so as to admit three fingers. After this the condition improved a little and the patient was discharged at her own request on Sept. 20th 1890.

Later History

After leaving the Hospital the woman became gradually /

gradually weaker and thinner with continuous incontinence of urine and faeces and recurring attacks of diarrhoea and vomiting. About the beginning of April 1901 an abscess gradually formed in the left groin and broke through the skin discharging thin pus. Soon afterwards a second abscess slowly formed at the upper and inner aspect of the left thigh. On May the 19th it is noted that the latter abscess had not yet broken. The ulceration round the urethra had spread.

There had been two menstrual periods in the last 10 months each lasting four or five days.

The patient died at length from exhaustion on May 23rd 1901 after several days of vomiting following on attacks of diarrhoea.

Observations.

1. The Case shows great hypertrophy, stricture of rectum and atresia of vagina ulceration of the vestibule and posterior vaginal wall.
   The ulceration in the groin in conjunction with the frequent attacks of diarrhoea and vomiting would suggest that the ulcerating process had affected the peritoneum and the rectal wall, causing irritation and subacute attacks of peritonitis.
2. Death was due to exhaustion; this is the usual cause recorded in severe cases of esthiomène. An attack of pelvic peritonitis may have directly led up to it as an additional factor.
3. The husband was a discharged soldier; the patient had never had any children. These two facts are the only ones available, pointing in the direction of luetic infection. The majority of cases on record which show such severe lesions as the case under discussion have a definite history of syphilis. Although this case does not show a clear history it is safe to assume on the strength of the two points given that the lesion was /

was one of tertiary syphilis.

4. Duration: apparently 11 years. Had the lesion been due to any other type of ulcerative process the patient would have succumbed much earlier. The long duration of the disease is typical of such tertiary lesions.

5. The absence of all response to anti syphilitic treatment is a fact frequently recorded with cases of esthiomène of this type. It is usually admitted that tertiary syphilitic lesions of the vagina vulva and anus do not respond to anti-syphilitic remedies. The fact that lesions in that locality do not respond, does not indicate that they are not syphilitic. Slight lesions frequently respond; severe lesions such as described as esthiomène perforant are known to respond only rarely.

V. 1906.

Case of Ellen Errington.

Age 58.

Personal History.

Patient has been married for 42 years and had 8 children; 4 died and 4 are well and strong. The youngest is 23 years old.

One miscarriage occurred 24 years ago.

Complaint: burning sensation in the region of womb and passage, constantly present; occasional sharp shooting pains in the vagina; some pain on micturition. There has been a bloodstained discharge for two years.

She has been getting thin the last two years and feels ill.

History of previous health.

Her husband is a soldier; she lived with him in India for seventeen years and there had low fever, "fever with ague " and small-pox. She has never been quite strong since.

Catamenia: Began at the age of 14, has always been regular and has no pain. Has always lost a good /

good deal, periods lasting 10 days.

Menopause at the age of 50.

Condition on admission:

Patient is very thin. Face flushed and rather drawn; has some pain.

Inspection of the vulva:

Round the urethra is a red fleshy body, very tender to the touch.

Abdomen: nothing to note.

Heart and Lungs: sound. Arteries are very much thickened. Vaginal examination revealed nothing abnormal with regard to pelvic organs.

Operation:

Patient was anaesthetised and carefully examined. A red fleshy mass was seen around the urethra and overhanging its orifice; it was sessile, the size of a green pea; it bled slightly when handled. Just inside the vaginal orifice on each side was a red patch not unlike a caruncle in appearance and around these bodies small red spots were visible. The cervix was dilated and the fundus thoroughly curetted; there was nothing abnormal in the appearance of the scrapings from the /

the uterus. The inside felt smooth. The mucous membrane of the red fleshy body around the urethra was incised and the body was dissected off. The raw surface was cauterised with Paquelin's Cautery. Picric acid solution was applied to the urea.

The mucous membrane over the patches in the vagina was incised and stripped off the vaginal walls: the raw surface was sutured with silk worm gut sutures; the vaginal wound was dressed with iodine ointment on lint.

Microscopic Examination:

Both the red patches excised from the vagina and the fleshy mass around the urethra show inflammatory tissue of great vascularity with extensive round celled infiltration and blood-extravasation. There are no new formed elements.

Observations.

1. This case resembles the condition which Huguier calls "Esthiomène superficiel, erythemaleux et tuberculeux;" "l'épaisseur de la peau semble être augmenté, excepte sur quelques parties. Çà et la on observe des points plus rouges, comme papuleux. Le tissu cellulaire sous - cutané est légèrement tuméfié et infiltré. Cette variété d'esthiomène coexiste le plus ordinairement avec les autres espèces d'esthiomène."

   The red raised patches in the vagina correspond to that description, while the fleshy mass around the urethra indicates that the process of hypertrophy of vulvar structures has begun.

2. The history in this case would point to syphilis (husband soldier in India) but there are no other points which support its assumption.

Ellen Errington.

(Low Power.)

This section shows the floor of the ulcer and the subjacent tissues.

The epidermis is completely disintegrated, showing only a few vestiges of oedematous epithelial cells.

The subcutaneous tissues are transformed into dense fibrous masses. In some places the connective tissue is completely organised forming a homogeneous mass; in other places strands of wavy thick fibres are visible.

The vessel walls are enormously thickened.

There is no vascular engorgement and no small cell infiltration; this denotes that the inflammatory stage is over and that the process of chronic fibrosis is completed.

Ellen Ericsson
(Low Power)

VI.

Patience Pearcey.

Aet. 38. April 17. 1905.

Esthiomène of vulva and vagina.

Present condition:

Vulva: Brownish red colour, somewhat swollen 'points' of shiny cicatricial nature. There is no ulceration of the labia majora. Two small external piles on the anterior aspect of the anus. On separating the labia minora widely a firm projection from the lower posterior vaginal wall becomes visible. It is of a pale red colour and moderately firm to touch.

On each side of the projection and just in front of the fourchette are two small ulcers each of about a ¼ inch in diameter. They are punched out ulcers with clearly cut edges covered by pale pinkish weak granulations. The depth of the right one is about ¼ inch.

The left runs obliquely upwards and backwards and communicates with the rectum on the anterior vaginal wall. Just behind the urethra are several flap-like valves moderately firm in consistence like hypertrophied caruncule myrtiformes. Their base is /

is situated on a very firm cicatricial looking mucous membrane. The mouth of the urethra is much contracted by this cicatricial mucous membrane and will scarcely admit a small catheter.

The vagina above this point is apparently healthy.

Vagina: The hypertrophic nodules extend half way up the vagina. Vagina is small and contracted.

Uterus and appendages are quite normal.

Personal History.

Married at 20.

Husband: bricklayer.

Children: five, eldest 19 years ago, youngest, 9 years ago.

Eldest died of croup.

Youngest died of inflammation of the bowels, five months.

All living, healthy, except the youngest.

The youngest has had a bad knee which was excised (tuberculous?)

Labours /

Labours all easy and normal with rapid convalescence.

Previous illnesses:

During carrying of last child, patient caught from husband a sore on the vulva but never had a sore throat or a rash. Carried child to full term and it was a healthy baby. After birth of the same child, six weeks after, patient had an abscess in the right groin, which took several weeks to come to a point. Doctor lanced it. It took several weeks to heal.

Family History:

Father died aet. 65. Cardiac disease.

Mother aet. 77, healthy.

Sisters, three alive and well.

No tuberculosis in family.

Present illness:

In January 1904 patient had rheumatic pains, and suddenly while sitting on a chamber she felt something burst between the anus and the vulvar orifice; this bled a little. Ever since that time she has always had some leaking from the rectum coming away into the vagina. Patient has no pain; her bowels are moved naturally and only leaks slightly after the bowels /

bowels have moved. She occasionally has a bearing down sensation and there is occasionally a little blood on the motion.

Patient has some difficulty in holding the water at night. Occasionally there is some increased frequency.

Menstruation:

Regular up to end of 1903. During 1904 & 1905 patient has only menstruated half the number of times; quantity normal.

Leucorrhoea for many years; occasionally blood-stained during last fourteen months. Bowels, regular. Appetite good. Not losing flesh. A scar in the right groin. No enlarged glands.

Observations.

1. This case resembles the case of C.N. Number 4. It corresponds to the type described by Huguier as esthiomène hypertrophique et perforant. A mass projects from the vagina. A vagina-rectal fistule is present and the urethral orifice is narrowed by a stricture. The features presented are usually found in the severe and advanced types such as they are described by the older French and English Writers, among the second chiefly Mathews Duncan.
2. There is in this case a clear history of syphilis; a definite infection causing a primary local sore and sterility since occurrence of the sore.

VII. 1902

Case of Olive Carter.

Patient was seen for the first time in the X Ray department by Dr Hall Edwards in November 1902.

Family History:

Patient is the only living child of a family of five. The first child died at the age of six, of spinal disease; the second died in infancy; two died since birth of patient, one of convulsions and one of pneumonia at the age of three. There were no miscarriages.

History:

Patient has always been delicate. Soon after an attack of chickenpox she began to feel pain and itching in the region of the vulva. This has now lasted for over fifteen months.

She was seen in February 1903 by Dr Th. Wilson who admitted her to Ward 15 of the General Hospital.

Condition on admission:

Patient is an intelligent looking little girl; she seems healthy and well nourished.

Tongue is clean; teeth badly formed; pulse regular; heart and lungs sound.

There /

There is a flat ulcer on the outer and upper third of the right thigh; its edges are sharply cut, wavy, brown. It has the appearance of a partially granulating ulcer. It has been treated with X Rays for several months. Its size is about that of a five shilling piece.

Two years ago there was a dark red raised patch on the left hip posteriorly, "which showed a slightly scaly surface similar to a crocodile's hide"; this patch began to ulcerate; it is now healed. There is a small naevus on the right lower abdomen. At the back of the right ear there is a small patch of lupus, almost completely healed.

Vulva: There is some redness of the vulva and lower part of vagina.

Two small flat raised patches with somewhat indurated bases are seen in the vagina. One is about the size of a French bean; it is situated on the right and posterior vaginal walls and invades also the anterior part of the perineum. The ulcer is quite pale; its edges are raised, irregular and thickened; the surface is irregular. The other ulcer which presents the same characteristics is about /

about the size of a split pea and is situated on the left vaginal wall.

Operation: The tuberculous ulcers situated in the vaginal wall and extending on to the perineum were dissected out. Patch of lupus on the right thigh was excised.

April 3rd 1903. Condition of the vulva:

Parts of former ulcerations healed.

A small granular irregular patch at the posterior part of the right labium majus and extending forward, is now visible; the surface is yellowish and uneven. Edges are only slightly raised and not well defined.

There is no induration of the base of this new patch.

October 21st 1903. New patch disappeared. Perfectly well.

Observations.

(1) Age of the Patient:

All the cases recorded as Esthiomène or lupus vulva in children appear to have been definite cases of lupus, (lupus in the sense of tuberculosis of the skin), with lupus patches elsewhere, or tuberculous lesions in bones, joints, etc., or tuberculous glands. The youngest patient I find on record as esthiomenic is the case published by J. Townsend in the Boston Medical and Surgical Journal, Sept. 1886. She was 15; her father was tuberculous. There was ulceration without hypertrophy. The ulcers described resemble those of Olive Carter; the case was in all probability one of tuberculosis.

(2) Absence of hypertrophy. An ulcerating condition without hypertrophy does not satisfy the requirements for a diagnosis of esthiomène. All the same numerous cases stand on record as esthiomène where there is no statement that hypertrophy existed.

The case is interesting in as much as it resembles many cases diagnosed as esthiomène or lupus /

lupus vulvae by the earlier writers, but is distinctly a case of tuberculous ulceration. This supports our view that esthiomène as a separate and special disease does not exist and that many diseases, such as syphilis, cancer, tuberculosis produce lesions which have been termed esthiomènic.

VIII. 1898.

Ulceration of the vulva of uncertain duration, with chronic pulmonary phthisis.

K. Monzer, widow aged 48 was admitted to the General Hospital Birmingham on June 28th 1898 with ulceration of the vulva.

Personal History.

She had had 2 children, the last 19 years ago and had been a widow for 15 years. There was evidence of chronic alcoholism.

History of present illness:

The patient's complaint was that she had been losing freely for three weeks before admission at irregular intervals with passage of clots. Previously the Catamenia had been regular.

Condition of the vulva.

There were two ulcers, one at the posterior commissure with its centre in the position of the hymen extending along the inner surface of the left labium and to some extent also on the right one; it also involved the lower end of the posterior wall of the vagina. The edges of this ulcer were irregular sharp and undermined. Surrounding it was a considerable /

considerable area of induration with ill defined edges of about $\frac{1}{16}$ in. in thickness. There was a moderate amount of thin brownish discharge from the depressed granular surface of the ulcer. The ulcer did not bleed, nor did the tissue in its base break down easily.

There was a second similar ulcer, the size of a shilling, on the inside of the left labium majus.

Extensive induration was present in the left labium majus and in the recto-vaginal septum. On June the 28th the ulcer was cleaned up under a general anaesthetic and a small triangular portion was excised for microscopical examination.

This showed that the base of the ulcer was formed of tuberculous tissue, most of it in the form of well-defined nodules, many with giant cells in their centres. In other parts of the section the tuberculous tissue had a more diffuse arrangement. The epithelium covering the surface of the section up to the margin of the ulcer showed no definite changes.

Condition on admission:

On admission the patient was very ill and considerably emaciated. She had a troublesome cough with /

muco-purulent expectoration which was said to have been present for some years off and on.

There were scattered bronchitic râles throughout the chest but no areas of consolidation.

The temperature reached 100° Fahr. on the evenings of the 24th and 26th June. Pulse was weak and compressible.

On June 28th the ulcer, together with the underlying indurated tissues was removed under a general anaesthetic. This was followed by a considerable increase of the chest symptoms, troublesome cough, muco-purulent expectoration and occasional attacks of shortness of breath. The evening temperature was 101° Fahr..

The sputum was twice examined for T.B. with negative results.

By July 20th 1898 the ulcer wound had soundly healed but the patient's general condition did not improve.

Patient left the hospital in July and died on the 24th August. Death was certified as due to pulmonary phthisis.

Mr Powell White examined the ulcer removed at operation /

operation and reported "the structure is typical of tubercle and tubercle bacilli can be obtained in situ".

Observations.

1. The patient was phthisical and died of phthisis pulmonale.
2. Cases of esthiomène or lupus vulvae occurring in people with phthisis may without hesitation be taken for tuberculous ulcerations. All such cases reported agree in showing that there is no hypertrophy, as in this case, or if some hypertrophy was present the ulceration was greatly in excess, and by far the most important feature of the condition. Usually such ulcers are reported as having undermined edges and of causing much pain.
3. The tissues were shown microscopically to consist of tuberculous tissue and the tubercle bacillus was found present.

In all cases of ulceration of the vulva with phthisis where a microscopic examination was made some element or elements pointing to tuberculosis were found. Older writers included such cases among their lists of "esthiomenic or lupoid" ulcerations, considering that they constituted one variety of that disease.

Table I.

Name and Status	Age	History of Syphilis	General Health.
I. Maggie Lawrence Married, uni-para. Press worker.	35	Yes	Delicate
II. Lucy Blair Married, uni-para. Wife of discharged soldier	28	Yes	Suffering from secondary syphilis
III. Ellen Hill Single	22	Probable	Not stated
IV. C. N. Married, multi-para. Wife of discharged soldier	24	Probable	Very poor. Profound cachexia.
V. Ellen Errington. Married, Multi-para. Wife of soldier in India	58	Probable	Poor
VI. Patience Pearcy Married, multi-para.	38	Yes	Poor

Table II.

Name	Symptoms	Diagnosis on admission.	Duration of condition
I. Maggie Lawrence	Pain and itching	Esthiomene	Three years
II. Lucy Blair	Discomfort on sitting and walking	Condylomatous Hyperplasia of anal and vulvar tissues	Two years
III. Ellen Hill	Not stated	Syphilitic and tubercular ulceration of the vulva	Three years
IV. C. N.	Incontinence of foeces and urine. Attacks of abdominal pains with vomiting and diarrhoea	Syphilitic ulceration	Eleven years.
V. Ellen Errington	Burning pain and dysuria	"Inflammation of the vulva."	Two years
VI. Patience Pearcy	Incontinence of foeces	Esthiomene	Two years

Table III.

Name	Condition of the Vulva.	Response to treatment
I. Maggie Lawrence	Marked hypertrophy of the clitoris and right labium minus; two shallow ulcers, some discharge. Enlarged gland in groin.	Operation: Excision. No after history.
II. Lucy Blair	Hypertrophied mass round anus. Stricture of rectum. Scar and gland in groin. Deep ulcer in right labium majus	Antisyphilitic treatment. Complete cure
III. Ellen Hill	Great hypertrophy of labium minus. Large ulcer on inner surface of right labium majus	Antisyphilitic treatment and excision cure.
IV. C. N.	Great hypertrophy of clitoris and labia majora. Strictures, atresia vaginal. Great deformity. Fistulae of rectum and urethra. Extensive ulceration.	Antisyphilitic treatment and operation. No response Death from cachexia.
V. Ellen Errington	Great hypertrophied mass round the urethra; two ulcers near ostium vaginae.	Cautery dressed with picric acid. No after history
VI. Patience Pearcy	Fistulous ulcerations extending from vulva into rectum: hypertrophied areas in vagina.	Operation and antisyphilitic treatment: no record of cure.

CHAPTER VI.

Part I.

Aetiology.

Age.

Esthiomène usually occurs between the ages of 20 and 55. Cases reported under 20 are generally tuberculous. There is no special age at which esthiomène is outstandingly frequent but it may be said that on the whole it occurs more frequently between 20 and 40 than later.

Pregnancies.

It is interesting to note that women afflicted with esthiomène rarely have more than one or two children. Absolute sterility, one child sterility abortions and premature births are common. The children are often reported to have been weakly and to have succumbed to some ailment in the first year of life.

Heredity.

There is no apparent relationship between heredity and esthiomène.

Social Status and Mode of Living.

There is no doubt that esthiomène is almost exclusively met with among the people of the lower social strata, the so-called "hospital class" of patients. The French writers collected their cases from among the large masses of destitute and often degenerate people who fill the wards of the large Parisian Infirmaries. In this country and in America the clinical material is also derived from the less fortunate section of the population. Esthiomène is more often seen in the wards of Lock Hospitals than in a General Hospital. In Great Britain it is comparatively rare and the very severe cases that die from exhaustion, such as the first French writers described are hardly ever met with.

Many authors consider that prostitution is the main aetiological factor. They lay much stress on the poverty, neglect, dirt, privations "la misere physiologique" (by which the French mean a marasmic state) which are usually attendant on that condition. This is not altogether true for esthiomène may occur in those who are clean in their habits and mode of living and who are not wasted. Other writers again point /

point out that blooming health is usually found and that the patients are remarkably robust considering the extensive ulcerations they are afflicted with. (Cases of Mathews Duncan and Dr. Grace Peckham Murray). As a matter of fact one view does not exclude the other. Early cases usually present a healthy appearance but the further the disease progresses the more the cachexia increases. In addition to this matter of degree there is also the question of constitutional disease such as phthisis or syphilitic lesions elsewhere. Amongst the cases collected by Dr. Grace Peckham Murray 21 enjoyed good health, 5 had poor health and 7 had no record regarding their general health.

Syphilis as an aetiological factor.

The most important of all aetiological agents in the history of a case of esthiomène is syphilis. The greater number of cases reported show a history of syphilis, either definite or probable. The relationship between syphilis and esthiomène will be discussed later.

Trauma as a predisposing factor.

Frequent local irritation producing inflammation warts and sometimes abscesses has been considered by many to be the sole cause of esthiomène. The Germans notably Veit and Schroeder have forwarded this view. There can be no doubt that constant irritation will increase the disturbance but it is not the determining cause.

Other local conditions are sometimes accepted as causes of esthiomène, such as hard and soft chancres, boils. These are rarely found present at the same time as esthiomène and though they may precede the latter they can hardly be said to be a local cause.

Theories that have been advanced regarding the pathological nature of esthiomène.

A rapid survey of the different views that have been held regarding the cause of esthiomène shows the difficulty encountered by the older writers to distinguish without the aid of the microscope between the various types of hypertrophy with ulceration. It must always be remembered that many of them wrote before the time of discovery of the Bacillus of Koch and the Spirochaeta of Schaudinn and also that many of them drew their conclusions from the patient's history and the clinical aspect of the case alone without verifying their theories by a careful examination of the tissues.

The following are the theories which have been supported by various authorities:-

Esthiomène is

(1) due to tuberculosis, local or general.

(2) due to syphilis a tertiary or a parasyphilitic manifestation.

(3) malignant disease either epithelioma or a form of rodent ulcer.

(4) a form of elephantiasis either a "Stauungs elephantiase" /

elephantiase" (lymphatic obstruction elephantiasis) or an "inflammatory elephantiasis, local thickening after inflammation.

(5) A purely inflammatory condition, most frequently caused by local irritation and trauma, sometimes following other lesions such as tuberculosis or syphilis.

(6) A disease sui-generis of unknown nature.

I.

Tuberculosis.

The idea that esthiomène or lupus vulvae is a tuberculous manifestation was very widely held. Its chief advocates among the eminent writers both of the 19th and the 20th centuries are Bernutz, Figuet and Pozzi. As we have already seen in the historical chapter of this work, Bernutz was the first who ascribed to "lupus of the vulva" a tuberculous nature. His view was adopted by Figuet whose theories on the "stroumous" nature of esthiomène have already been given and who saw a connection between pulmonary phthisis and "lupus vulvae" in a third of his cases. Finally Pozzi in 1907 forwarded that he thought all cases of "lupus vulvae or esthiomène" were either directly /

directly or indirectly tuberculous.

Comparatively few of those who retained the term of lupus and who saw a resemblance between lupus vulvae and lupus facialis considered that they were of the same pathological nature. It was in the days before the nature of lupus facialis was known and therefore the term lupus did not convey any more definite meaning to those who used it than that it implied a process of eating away. In that sense also the term was used in connection with ulcerous hypertrophy of vulvar structures. Many writers ventured no opinion regarding the cause of the latter and merely retained the term lupus on account of a superficial resemblance between esthiomènic ulceration and lupus facialis. Huguier, Guibout, Angus Macdonald and Mathews Duncan called esthiomène lupus and saw in it a resemblance to lupus facialis but did not accept it as a tuberculous manifestation. Huguier's essay begins as follows: "Cette maladie chronique qui tient le milieu entre l'éléphantiase des Arabes, la syphilis le cancer et la scrofule etc etc " Guibout says: "Il y a réellement à la vulve qu'à la face un esthiomène" He makes no statement as to his views regarding the nature /

nature of this "special disease."

M. Duncan makes the following statement:- "the name of lupus is retained in order to avoid change and because the character of the disease brings it into alliance with the ordinary run of such cases." He uses the term lupus in preference to esthiomène since it expresses the great eroding character of the disease, including ulceration inflammation and hypertrophy variously combined. In his first essay "Lupus of the Pudendun" (Med. Times 1884) he states that: "there is much resemblance between this lupus and that of other parts but I do not tell you that they are pathologically the same as is generally believed." And finally he concludes by saying: "the disease has been said to have alliance with scrofula but I have failed to trace clinically any such connection."

In the same strain writes Angus Macdonald. From him we have this statement: "there is no essential difference in lupus of the vulva from lupus of any other part. Various attempts have been made to connect lupus with certain cachectic conditions such as syphilis or scrofula. The latter view is especially common among the French authors."

From /

From the foregoing we conclude:

1. That to many authors lupus was a term which implied a slowly increasing and widely destructive ulceration the nature and cause of which were unknown.

2. That owing to a superficial resemblance between lupus on any part of the body and esthiomène the name of the lupus was applied to the latter. Before Huguier's introduction of the name of esthiomène all ulcerations of the vulva were called lupus. He did not discard the old name and much confusion resulted especially later when the nature of lupus became known.

3. Some of the older writers saw a relationship between lupus and scrofula and there were some amongst them who adhered to the name of lupus in speaking of esthiomène and held that it was of a tuberculous nature. Others again, aware of the relationship of lupus to tuberculosis, rejected the view, that esthiomène was a tuberculous lesion but still did not discard the name of lupus when describing cases of esthiomène.

4. The name of lupus since in the modern meaning it signifies /

signifies a tuberculous lesion of the skin and subcutaneous tissues is utterly fallacious as applied to cases of esthiomène for the latter is not tuberculosis of the vulva. Vulvar tuberculosis presents totally different characteristics from esthiomène as we shall see when discussing the differential diagnosis. The term lupus should therefore be omitted altogether in discussion of esthiomène and in order to avoid confusion it might be better to use the term "Tuberculous ulceration of the Vulva" in those cases where tuberculous tissue is found under the microscope.

II.

The syphilitic nature of esthiomène has been admitted by many, notably Jonathan Hutchinson, Malcolm Morris and the German authors. It is decidedly surprising that many writers who published exhausting monographs on the subject should have omitted to enquire more carefully into the histories and nature of other ailments of their patients. In many cases there is no record as to whether there has been a syphilitic infection or not, whether there were any lesions present apart from the vulvar condition indicative of syphilis. In Dr. Grace Peckham Murray's tabulated /

tabulated analysis of 33 cases we note that 19 probably had had syphilis, 7 had a distinct history of syphilis and the rest showed no record. A general survey of the literature of the cases published as esthiomène, with a careful study of the histories and clinical conditions of the cases impresses the mind strongly with the fact that the one and only cause of esthiomène is syphilis. We shall discuss more fully in a later chapter the connection between the two. It suffices here to say that amongst the authors of monographs on esthiomène Dr. Grace P. Murray and Deschamps admitted that besides cancer and tuberculosis syphilis is another causal factor in the production of esthiomène; in other words they made syphilis responsible for some of the esthiomenic ulcerations. Veit, Koch, V. Winkel and other German writers of monographs on esthiomène are alone in declaring syphilis to be the only cause of esthiomène.

III.

Dr. Grace Peckham Murray and Deschamps made cancer another occasional cause of esthiomène. They held that a certain number of cases were malignant. Brodie Cooper and Paget thought all cases of esthiomène /

esthiomène were malignant. Macnaughton Jones held that "lupus vulvae may be considered an epithelioma" and again he said that it might be considered in the light of a rodent ulcer. He did not publish any microscopic examinations to support this view. There is no doubt that malignant degeneration sometimes occurs (as in the case of Maggie Lawrence). The cancerous growth may increase so rapidly as to destroy all the characters of preexisting esthiomène. The occurence of malignant degeneration has probably contributed to the reputation of mortality of the disease.

IV.

Esthiomène or lupus vulvae is another form of elephantiasis according to the views of most German authors who discoursed on what they called Ulcus Vulvae. The term "Stauungs elephantiase" was introduced by F. Koch by which he meant elephantiasis due to lymphatic obstruction. This lymphatic obstruction he thought was either due to previous extensive lymph glandular inflammation, with subsequent suppuration or removal. His idea was that after such an occurrence (usually due to syphilis) the powers of resistence of the vulvar tissues against trauma suppuration /

suppuration and diseases such as tuberculosis and cancer. He was unable to prove either syphilis or tuberculosis as causes in his cases. Other authors shared Koch's view that esthiomène was a form of elephantiasis though not necessarily due to obstruction of the lymphatics, they considered it in the light of an elephantiasic connective tissue hyperplasia resulting from frequent superadded attacks of inflammation in the tissues producing a resultant overgrowth.

V.

Esthiomène is a purely inflammatory condition most frequently caused by local irritation and trauma, sometimes following other lesions such as tuberculosis or syphilis.

That it is a purely local and inflammatory condition without relationship to any constitutional disease is a view which we frequently find expressed. Andre Boursier (in the Précis de Gynécologie) held that esthiomène "est une ulcération chronique de la vulve due ordinairement à des causes banales et se compliquant d'un élément hypertrophique résultant d'un processus éléphantiasique."

Again /

Again Dubreuilh et Bran in their Thèse de Bordeaux stated that it is merely "une ulcération chronique ne présentant rien de specifique."

Finally Dupuy et Rullier maintained that it is neither syphilitic nor tuberculous in origin but merely a chronic ulceration with subsequent hardening; they termed it a scleroma.

R. W. Taylor (1889) thought that some cases were due to an inflammation after trauma, irritation and other mechanical local causes, while others followed on syphilitic infection but not being syphilitic manifestations themselves. He states: "that a large and perhaps greater number of chronic deforming vulvar affections are due to simple hyperplasia of the tissue induced by irritating causes, traumatisms and inflammations;" "that many cases are due to simple hyperplasia in old syphilitic subjects who suffer from chronic ulcerations of the vulva long after all specific lesions have departed."

VI.

The last view to be mentioned and the one least frequently met with is that esthiomène is a disease per se or sui generis of unknown nature. We have already /

already seen that Isaac Taylor of New York was the first writer who advocated that "esthiomène or lupus might from several peculiarities which appertain to it be considered a disease sui generis." He did not forward any theories as to the nature of this special disease, but was inclined to think it"a local disease grafted on a constitutional one such as cancer, syphilis or tuberculosis!" He has apparently remained the only advocate of this view.

Part II.

The Author's Views on the Nature of Esthiomène.

A careful analysis of the cases of esthiomène that we were enabled to study, and a perusal of the large mass of literature published on the subject lead us to the conclusion that it is a tertiary manifestation of syphilis and that the syphilitic virus is the only cause of the condition. There can be no doubt that a number of accessory factors have a marked influence on it but they could not be the cause of a state which, considered from every possible point of view bears the stamp of syphilis. Such accessory causes are constant local irritation with its attendant results of inflammation and sometimes abscesses and warts, uncleanliness and above all a low state of health aptly termed by the French "la misère physiologique." That irritation and inflammation of the parts could be the sole causative agents of a state characterised by deep and widespreading ulceration extending into the pelvic organs, as some writers considered is inconceivable. Nor can we understand how modern writers were able to read "tuberculosis" into a case of esthiomène, manifestly so /

so different in appearance, histology, history, symptoms and results. We append two cases of tuberculosis of the vulva to show the difference between the two conditions. A case of esthiomène is occasionally met with which may at first simulate tuberculosis or cancer or an indurative elephantiasic oedema. Such cases are however rare and a careful enquiry into the history cannot fail to enlighten the student regarding the class of disease to which such a case should be relegated. Where a portion of tissue can be removed for examination a first glance through the microscope reveals the true nature. With our present day knowledge of the bacillus of Koch the spirochaeta pallida, the tissues of gumma, cancer, etc. and rapid histological methods no mistakes of diagnosis should arise. It is interesting to note that in recent years the term "lupus vulvae" has given way to the name of "tuberculosis of the vulva" and that the non-committal name of ulcus vulvae has disappeared. Huguier's term "esthiomène" is now but rarely used which seems to us a matter of regret for if it is used in the definitely understood sense that it is a tertiary syphilitic manifestation it replaces usefully /

usefully the cumbersome expression: "syphilitic hypertrophy of the vulva with ulcerations."

We shall now proceed to state on what grounds we base our conclusion that esthiomène is a tertiary syphilitic lesion.

History.

The history has already been fully discussed. The points we wish to recapitulate here are the following:-

Social Status.

The poor and neglected furnish the largest number of cases; the unmarried are in excess of the married though that fact in itself is not of great importance. Servants and workers in laundries among the unmarried and wives of soldiers and sailors among the married are frequently the victims of esthiomene. Sterility, relative and absolute is often recorded. Large families are only found where esthiomene appeared later in life.

Age and period of occurrence.

"As regards the date of the outbreak it can be said that tertiary syphilis may appear at any date after infection from the first year onwards; in rare cases they may become manifest even while the secondary eruptions are still present. More usually there has been a period of quiescence and apparent health between the secondary and tertiary phenomena; years of greatest incidence are the second and /

and especially the third after infection." (F. W. Andrews). The variability in time of early tertiary lesions, which may appear early or very late, at the end of the secondary period or after a long time of apparent cure explain the great differences in the age incidence of esthiomène. Although from 20 - 40 seems the most usual period for its occurrence it may be present earlier or later. There is in fact no age and no period at which it may be said to occur with special frequency. It is often a lesion indicative of latent syphilis, a sudden granuloma appearing in the region of the perineum after many years of apparently blooming health. Of the appended cases we have give the following histories bearing on syphilis:

1. C. N. aet. 24, wife of a discharged soldier, never had any children.
2. Maggie Lawrence, aet.35,Pressworker. Married at 18, had two abortions in the first year of marriage,one at the end of the second month and one at the sixth week. In the second year had a fullterm child who died in its first year. No children since though married 17 years. "After marriage began to /

to feel ill and has never been quite well since."

3. Ellen Errington aet. 58. Patient has been married 43 years and had 8 children 4 of whom died. Her husband is a soldier; she was in India with him for 17 years.
4. Patience Pearcy aet. 38. Married at 20. Had 5 children. "During last pregnancy caught a sore on the vulva from the husband." After birth of child had an abscess in the groin which was lanced and took a very long time to heal. Youngest child is 9 years.
5. Ellen Jones Hill aet. 22, single. Three years ago had a "breaking out" on the vulva.
6. Lucy Blair aet. 28. Married at 20. One child born 3 months after marriage lived 2 days. No pregnancy since. "She has had bad sore throats several times;" has a rash on the arms from time to time and suffers from severe headaches. Husband has been a brasscaster and hawker alternately since the age of 15. Five years ago he was in the army for 1½ years but was discharged.

Conditions of General Health.

For many years esthiomene does not alter the state of health markedly, though the disease may be extensive. This is a distinguishing feature from tuberculosis or cancer which we shall discuss in a later chapter. It cannot however be said that the patients are blooming in health and robust in appearance; there is almost invariably something else the matter with them, recurrent rashes, headaches, lassitude or some degree of anaemia. The patients whose cases we are studying in this thesis were anything but healthy. Where there is fistulas burrowing with incontinence the condition of health is very distressing. There is often a marked degree of cachexia even at an early stage of the disease. The fact remains however that there is no immediate attack on the general health of the patient who may go about doing her work for many years with large ulcerations without much inconvenience. Examples taken from among the cases studied:

1. C. N: emaciated and anxious looking. (Severe case.)
2. Maggie Lawrence: Patient is thin and spare and looks /

looks delicate.

3. Ellen Errington: Has been getting thin the last 2 years and feels ill.

We may accept it as a rule that where an esthiomène appears soon after infection where there are still some signs of the constitutional disturbance of the second stage, such as febrile and anaemic manifestations the condition of health is of necessity poor, but where esthiomène is an indication of "latent syphilis" coming on, often as an isolated manifestation of tertiary syphilis after many years of apparent cure of the disease, the general health is as a rule excellent. In the former class of case there is often immediate reponse to treatment, to the latter no reponse. It is the cases of the second kind that are apt after many years of ulceration to burrow deeply and to kill the patient by degrees through exhaustion.

Symptoms.

It is a matter of constant surprise to find how little inconvenience the esthiomenic state of the vulva causes. Some patients apply for medical relief at once because they are alarmed by finding an ulcerous or hypertrophied condition; others do not seek advice until a troublesome discharge or some itching causes inconvenience; others again come when the ulcers have already attacked the urethra bladder or rectum to be treated for incontinence. These are however rare. There is rarely any pain, darting or burning in the parts and they are handled without discomfort to the patient. We are well acquainted with the painless character of gummatous and other syphilitic ulcerations which helps us so often to distinguish them from tuberculous or inflammatory ulcerations.

Enlarged Inguinal Glands.

The syphilitic bubo is an occurrence of the first stage of the disease. Its presence or absence in esthiomène neither helps nor discounts the diagnosis. Where enlarged glands are reported it is frequently the case that esthiomène has supervened at a very early stage on the primary and secondary lesions. Enlargement of glands due to non-specific infiltration sometimes occurs in those who have suffered from those ulcerations of the vulva which have become "mixed". The hard small buboes with stringy thickening of lymphatic vessels between the beady glands are so typical of early specific diseases that a "septic" gland, large isolated and not nearly so hard as the bubo is readily distinguished from the former. Enlarged glands are rarely found in esthiomène.

Cicatricial tissue in tertiary syphilis.

The fibrosis that occurs around necrosing gummata is partly due to the reaction of surrounding tissues to the virus and partly to the fact that the gumma has broken down in the centre only and has left its spreading margins intact. For the fibrous process will spread in one direction while it is checked by necrosis in another. It has a tendency to step into the breach made by the ulcerative process and to repair the broken surfaces and spaces. It is however itself liable under renewed attacks on the part of the virus to break down and ulcerate. Another characteristic it possesses is its quality to contract. The presence of large masses of scar tissue therefore predisposes to great deformity. This is seen in esthiomène of many years standing. The parts are often quite unrecognisable. The strictures they produce are very difficult to deal with and persist when all traces of ulceration and hypertrophy have disappeared.

Duration. Course and Causes of Death.

The chronicity of esthiomène is a significant feature. All ulcers of a tertiary syphilitic nature are notoriously slow to progress and slow to heal. In no other ulcerative condition known, is the history so prolonged, so eventful of recurrences, so little disturbing in its effects. The course depends largely on the early recognition of the condition and the nature of the treatment enforced. It may be a matter of only a few months or it may last a great many years in which case death almost invariably ensues.

The causes of death are significant of syphilis. The syphilitic virus kills by indirect methods; it causes a general destruction of tissues until finally one or other system specially hard hit refuses to function and dissolution results; for example, if the arterial system is especially affected we have aneurismal dilations and subsequent rupture. Likewise with esthiomène, the ulceration itself would be of no importance from the point of view of causing death if it were not for the fact that the fistulae it produces determine the onset of wasting from malnutrition /

malnutrition. Copious thin foecal material is constantly escaping through the recto vaginal aperture or it may be through a communication between the vagina and the small intestine in which case wasting is even more rapid. The ulceration, burrowing widely may affect all the pelvic viscera and finally reach the pelvic peritoneum and establish a fistula between the viscera and the pelvic peritoneal cavity. This was the case in C. N. who suffered from numerous attacks of severe abdominal pain accompanied by vomiting and diarrhoea, no doubt due to intermittent slight attacks of pelvic peritonitis. These add to the other causes of cachexia to which the patient finally succumbs.

Response to treatment.

It has been said that esthiomène cannot be a syphilitic lesion because it does not respond to anti-syphilitic treatment. Anyone who is acquainted with tertiary lesions of that disease is aware that many of them do not respond to mercury and potassium iodide. It is interesting to note that syphilitic manifestations situated at the beginning and at the end of the alimentary canal remain unaffected by them. Leukoplakia of the tongue and syphilitic strictures of the rectum are specially noted for their indifference to mercury and potassium iodide; esthiomène may be added to these. It cannot be said however that it always fails to answer for the majority of early cases subjected to these remedies recover completely. But when the disease has lasted for some time and has been left untreated or been treated surgically only or with other remedies then the response may be slight or nil. Advanced cases with fistulae and strictures do not respond in the slightest degree. Authors have been in the habit of calling a case syphilitic or non-syphilitic according to the positive or negative results of anti-syphilitic treatment.

This /

This is obviously erroneous for the reason already given, that many tertiary syphilitic lesions do not yield to the treatment and secondly also because potassium iodide has an alterative influence on inflammatory conditions other than syphilis; thirdly, if accessory lines of treatment, rest, feeding, cleanliness, scraping of the ulcer are added to antisyphilitic treatment it would be obviously fallacious to ascribe the cure to the latter alone. This is however often done. Amongst our own cases we note that among early occurrences cure was obtained by anti syphilitic treatment alone. (Lucy Blair) On the other hand the advanced and severe case of C. N. in whose case antisyphilitic treatment was carried out intermittently over many years there was no response and the patient died from the results of deep seated extensive pelvic ulcerations. In the other cases antisyphilitic treatment and operative interference by the excision of the hypertrophied masses effected cures.

The earlier the appearance of esthiomene after the primary infection the better the results to treatment.

Histology.

It is customary to regard the formation of gummata and the process of fibrosis, which are the essential lesions of tertiary syphilis, as two distinct changes. This is not strictly the case. Both are new formations of connective tissue and at their outset are similar; but whereas in fibrosis permanent fibrous tissue is formed, in gumma the cells tend to early necrosis and death. In the one case the reaction achieves its goal, in the other it fails. If this be so, it is no matter for surprise that the two processes are often seen in company, i.e. that failure occurs in one part of an affected area and success elsewhere. (F. W. Andrews in "A System of Syphilis" edited by d'Arcy Power and I. Keoch Murphy.) Have we not here the exact picture of what actually happens in esthiomène? Is not the condition one of constant fibrous tissue formation, showing an ever-spreading margin of new granulation tissue, a heaping up of old fibrous tissue with a tendency to necrosis in one part and cicatrisation in another?

That the hypertrophied masses are gummatous and consequently undergo the common fate of gummata in breaking /

breaking down into necrosing tissue leaving indolent chronic slow spreading and slow healing ulcers, of that can be no doubt. The cell elements are the same as those seen in gummata, the inflammatory cells are mostly plasma cells and lymphocytes. Numerous capillaries are present; the fibrous coats of vessels are enormously thickened at an early stage. When the gumma is somewhat older the intima in time also hypertrophies. At any period of the existence of the gumma central necrosis may take place. It may occur when the tissues are still infiltrated by thick masses of plasma cells and the gumma may as yet be composed of young granulation tissue; or it may occur later when the well formed connective tissue has already settled down into strands of wavy fibrillated bundles. When necrosis begins there is first of all a loss of all traces of cell staining. The structures look uniformly granular or gelatinous. A zone of small darkly staining cells surround this early necrotic tissue and at the edge the never-failing fibrous tissue ring is preserved from which cicatrising process spread inwards when the necrosis and discharge of broken-down tissue are completed.

In /

In going over the section relating to our cases we find all the phenomena as indicated above regarding gumma. There are numerous areas where fat deposits, degenerating cells and absence of cell elements mark the early stages of the developing ulcer. They are surrounded by the ring of small round lymph cells and plasma cells. At the edges of the hypertrophied masses we note the presence of the intact connective tissue from which cicatrisation would have proceeded.

It may be asked "if esthiomène is of a gummatous nature with slow fibrosis and necrosis and repair, how can one account for those cases of rapid deep ulceration, spreading from the vulva into the pelvic organs and sometimes causing death within a few months and sometimes after many years of renewed attempts at cicatrisation"? (One case reported by Munier died 15 months after onset of esthiomène.) We give the answer in the words of F.W. Andrews: "In addition to the formation of local gummata there is a diffuse type of perishable granulomatous infiltration, which may attack the skin but is also apt to affect the fauces pharynx and larynx. Here the tissues seem to melt away as it were by a process of rapidly /

rapidly spreading ulceration. The loss of tissue may be very great and the cicatricial contraction of the tissues concerned in repair may lead to serious deformities."

Granulomatous infiltration with wide spread ulceration appears to occur in the ill nourished wasting people with no powers of resistance. It appears to have some analogy to phagedenic ulceration which undoubtedly occurs in those whose tissues from some cause or other show no attempt to combat the syphilitic virus.

It may be stated here, incidentally that phagoedena is a rare occurrence in tertiary syphilitic affections of the vulva. The granulomatous infiltration described above has often been considered as phagoedena, it is however not as rapid and as destructive as phagoedenic ulcerations.

Why is Esthiomène a tertiary Manifestation of Syphilis?

1. Histologically esthiomenic tissues are gummatous. They undergo the usual processes of necrosis and ultimate cicatrisation with contraction that we observe in gummata.
2. Esthiomène usually makes its appearance several years after the primary infection.
3. There rarely are concomitant lesions pointing to secondary manifestations. Sometimes a "rash" is present and occasionally recurrent sore throats are found but on the whole the appearance and subjective phenomena point to the third stage. Headaches and pain in the bones are fairly common.
4. It is a wellknown fact that ulcerations and hypertrophies of secondary syphilis yield much more readily to treatment than those of the third stage. We have noted that esthiomène is obdurate to treatment in a large number of cases.
5. Professor Fournier shows that the skin is the commonest site for early tertiary lesions. It is not surprising therefore that in esthiomène we /

we often see no other indications of tertiary syphilis. This absence of other lesions may account for the fact that so many writers on esthiomène mistook its nature.

Summary of Chapter on Aetiology.

1. Esthiomène is a tertiary syphilitic manifestation, because:-
   (a) a history, direct or probable of previous syphilitic infection is almost always obtained.
   (b) the symptoms, the chronic course with relapses, and the indirect modes of producing death indicate syphilis.
   (c) a large number of cases respond to antisyphilitic remedies.
   (d) the Morphology is analogous to tertiary lesions, the gumma with its necrosis and gradual spreading ulceration with subsequent cicatrisation is typical of syphilis. In no other diseases do we obtain an active hyperplasia combined with an active ulceration.
2. Esthiomène is not a disease per se.
3. It has only an occasional relationship to carcinoma; it may undergo malignant degeneration.
4. It has no relationship to the bacillus of Koch.
5. It is not due to lymphatic obstruction, i.e. it is not a form of elephantiasis.

CHAPTER VII.

Differential Diagnosis.

Part I.

(a) Tuberculous Ulceration of the Vulva.

(b) True Tuberculous Hypertrophy without Ulceration.

(a) Tuberculous Ulceration of the Vulva.

This is a comparatively rare condition. H. Kelly in his Operative Gynaecology states "that tuberculous disease of the external genitals is extremely rare, not more than three or four cases having been recorded." He himself had only seen one case.

We append two cases of tuberculous ulcerations of the vulva to this thesis. Both had signs of tuberculous disease elsewhere; K. Monzer succumbed to phthisis pulmonale one month after her discharge from hospital and Olive Carter suffered from multiple lesions of lupus vulgaris.

The main diagnostic points between the vulvar ulcerations due to tuberculosis and those due to esthiomène.

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1. In tuberculous ulcerations of the vulva we always find a tuberculous lesion elsewhere, for it is never primary.

The primary lesions are most commonly in the lungs and the lymphatic glands, frequently in the skin, the peritoneum and intestines, sometimes in the renal organ the internal organs of generation. In children we find frequently tuberculous lesions of lymph glands, in adults the commonest lesion is phthisis.

Figuet's cases almost all had " a strumous history" and cavities were found in the lungs of his patients after death.

X. Bender collected a series of 32 cases of tuberculosis of the vulva (Revue de Gynécologie et de Chirurgie Abdominale Sept. Oct. 1906.) All the cases were secondary manifestations of tubercle in other parts.

2. The age of the patient should be noted. Esthiomène does /

does not occur in children because it never results from congenital syphilis. It does not occur under the age of 20, at any rate it has never been reported in adolescents under 20. Ulcerations of the vulva occurring in children are either inflammatory or tuberculous. When there is a history of lupus or enlarged glands and when it has persisted for some time tuberculosis is indicated. Eight of the 32 cases collected by X. Bender were children; the others were adults over 40.

3. The symptoms of vulvar tuberculosis are much more severe from the beginning to the end of the disease than those of esthiomene. As soon as the ulcer appears, sometimes even before then, when there is only a small pimple with an unbroken surface, the itching and pain begin. These increase with the spreading of the ulcer. The symptoms complained of are intense and incessant burning and itching of the parts which demand scratching, and rubbing, great pain on and after induration when the urine comes in contact with the ulcerated surface; often there is lancinating pain in the vulva shooting down the thighs and into the pelvis.

4. /

4. Condition of general health is usually reported as very poor in those that have phthisis concurrently. It would appear therefore that vulvar tuberculosis is a fairly late affection in phthisis. When there is lupus or where the glands have been enlarged the general state of health may be quite good.

5. The chief feature that distinguishes tuberculous from esthiomenic ulcerations is the absence of hypertrophy. We have mentioned three cases where great hypertrophy of the whole of the parts was present, so as to simulate sarcoma condyloma or esthiomene but there was no ulceration present. When there is ulceration the hypertrophy is absent. Polypoidal growths and warty nodules have been observed in cases where hypertrophy existed. Without ulceration many cases show an inflammatory induration and enlargement of the parts affected by tuberculous ulceration; they are usually brawny red and hard and only temporarily increased in size. Such enlargements are not "hypertrophies" the parts thus enlarged are usually acutely painful and very tender to the touch /

touch.

6. The ulcers differ in all their characteristics from esthiomenic ulcerations. They are usually situated on the labia, commonly the minor aspect, and sometimes in the vaginal walls. Kelly's case shows a triangular ulceration around the urethra which is an uncommon site. The ulcer is usually flat, and tends to spread in extent rather than in depth. It is irregular in outline and the edges are as a rule quite uneven. They are sometimes thick rounded red and angry looking; more often they are thin, toothed jagged and undermined, with unhealthy grey granulations. The floor of the ulcer is usually shallow and slightly depressed below the surface of the surrounding tissues, sometimes it is somewhat raised. Its surface may be uneven presenting a tuberculated appearance with greyish white small points, but it is more usual to find a thick greyish white slough covering up the whole of the floor. When the slough is removed, the floor is usually found covered by yellowish granulations. The floor margins and immediately surrounding tissues may /

may be indurated. Often we find a brownish serous discharge from the ulcer but it does not bleed nor break down freely; caseation is uncommon but has been reported. It is sensitive to the touch.

7. Course and Results.

If untreated the tuberculous ulcers will spread in extent in the manner characteristic of lupus. It may heal in parts and then proceed again in an intermittent fashion. In a robust patient it may heal itself; in those afflicted with late phthisis it is likely to end only when the latter disease causes death. Tuberculous ulcerations may cause death in delicate subjects and seem especially fatal in children. But the patient succumbs more frequently to the primary lesion. There is no tendency to fistulous burrowing, strictures and deformity so characteristic of esthiomene, because there is less destruction in depth than in extent. While esthiomene remains as a rule limited to the perineum and chiefly the vulva, tuberculous ulcers, often multiple may by extension and confluence cover the adjacent parts of the perineum.

8. /

8. Response to treatment.

In early cases, youthful subjects with strong constitutions and few tuberculous lesions the prognosis after complete excision is very good. The application of Finsen Light appears not to be effective, possibly because where they were tried the ulcers may have been situated in the vagina and were not directly exposed to the light.

Early and complete excision with some of the surrounding tissue on the whole part of the vulva if the process has extended over its entire surface is the best and most rapid mode of producing healing. Where there is phthisis of fairly late date, surgical interference is not advocated.

It is unnecessary to await the results of treatment in order to establish a diagnosis; should the difficulties in deciding between esthiomene and tuberculosis be very great, which, we hold could only occur in an early case of either condition a course of antisyphilitic treatment will show the nature of the ulceration. This will influence the former while the latter will remain unaffected. Where operative means are resorted /

resorted to from the outset, one may experience a recurrence of either state but the greater likelihood of return is in esthiomène. A course of mercurials with potassium iodide will then clinch the diagnosis.

9. The Histology.

A small portion of a hypertrophied area or of the edge removed from a tuberculous ulcer and examined microscopically will reveal the usual tubercle foci with the bacillus of Koch present.

The examination of the wedge of tissue removed from the ulcer of K. Monzer revealed "that the structure is typical of tubercle and tubercle bacilli can be stained insitu." In all ulcerations of slow unyielding character, obviously not due to mere trauma or abscess, a microscopical examination settles its nature.

(b) True Tuberculous Hypertrophy without Ulceration.

Five cases of this extremely rare condition have been recorded. They all show marked hypertrophy of tissue without a trace of ulceration and were diagnosed as sarcomatous growths before removal.

Poeverlein of Munich reported a case in Hegars Beiträge Band III. Part I. of a woman 49 years old showing no signs of syphilis. She had a tumour the size of a five shilling piece on the inner aspect of the left labium minus with an uneven discharging surface. A diagnosis was made of sarcoma but the microscope revealed typical tuberculous tissue. There were no ulcerations.

Petit and Bender of Paris in his Revue de Gynécologie et de Chirurgie Abdominale, Nov. et Dec. 1903 Part VI. report the case of a woman aged 31 with elephantoid growth involving the labia majora and minora. There was no ulceration.

M. M. Forgue and Massabau, and Boursier published cases of tuberculous hypertrophy in 1909 and 1908. The first authors called their case "tuberculous elephantiasis." The structure showed oedema of /

of the corium, absence of sebaceous glands and numerous deep seated tubercle foci with small celled infiltration around them. Some tubercle bacilli were stained insitu.

Another case showing the same characters was reported by Meriel of Toulouse: Annales de Gynécologie et d'Obstetriques Dec. 1909. These five cases show that besides the purely elephantiasic enlargement caused indirectly by tuberculous infiltration and impairment of lymph glands and constituting the so-called "tuberculous elephantiasis" there is a tuberculous hypertrophy simulating elephantiasis. The latter is caused by the presence of tubercle bacillus in the tissues, the former by lymph stasis. The two can only be distinguished from each other by a careful microscopic examination. We hold that the epithet "tuberculous" prefixed to the term elephantiasis where there is no tuberculous disease present in the hypertrophied tissues is misleading and should therefore be omitted.

Tuberculous hypertrophy without ulceration can be distinguished from esthiomène by the same characteristics that distinguish all uniform enlargements without /

without ulceration from esthiomène.

These will be discussed under the heading of elephantiasis.

The best mode of obtaining a certain knowledge of the nature of such hypertrophies is the careful examination of the tissues under the microscope.

Part II.

Malignant Disease.

Carcinoma of the Vulva and Esthiomène.

There are a number of salient characteristics which at a glance distinguish the two conditions from each other.

The common age for carcinoma is 45 - 60. Social status and habits of life have no part in predisposing to it.

The sites most commonly selected are the clitoris and the labia majora while in esthiomène the latter are rarely and then but late affected. From the outset of the disease and often before it has actually revealed itself there is intense itching and burning with darting and stabbing pains in the vulva. Owing to this the patients often seek advice early and it is then possible to interfere effectively.

As to the course and result there appears to be a divergence of opinion regarding carcinoma vulvae. We speak here of the squamous celled carcinoma or epithelioma which is the common form of malignant disease in the vulva. Many writers hold that it is indolent /

indolent slowgrowing slowspreading and of low malignancy; that an epithelioma may be present for many months and even years without giving rise to much discomfort and without spread or gland infiltration. Others again hold that it is rapidly spreading and very malignant with early gland infiltration and a very grave prognosis. This difference of opinion is probably determined by the fact that there are two main forms distinguishable; one is the papillary form with rapidly ulcerating surface, the other the button like epithelioma with slow central ulceration and gradual breaking down into a crateri form ulcer. (Orthmann.)

In both forms carcinoma differs in all its features from esthiomène in its beginning course and ending. No preliminary irregular hypertrophy of parts takes place with subsequent ulceration, healing and renewed breaking down. A localised subcutaneous sometimes round sometimes lobulated thickening takes place which reaches the surface and then breaks down. Later a fetid saneous discharge flows from it; new nodules appear in the vicinity; the surrounding skin is brawny and unhealthy. When the /

the inguinal glands are enlarged the sore in the vulva assumes a choked brawny elephantiasic appearance c an irregularly convoluted surface. (Kelly.)

While in esthiomène the hypertrophy of the parts is quite irregular and the outlines are soon lost, a carcinomatous growth preserves the shape of the part until such a time when the whole part sloughs and fungates. In the matter of preservation of the form of the part it resembles elephantiasis.

When the ulceration has become widespread a mixed septic process supervenes which accelerates the breaking down process. It is interesting to note that in the esthiomenic ulcerations a septic process only sets in late; it does not assume the severe forms that carcinoma does. Sloughs and pus are not often recorded but they do occur when the ulceration is deep and the destruction of parts enormous. The absence of discharge, fetor and fungating tissue constitute one of the diagnostic features of the condition.

The constitutional symptoms are throughout the course of the disease much more severe than those of esthiomène. It is true that in some cases of epithelioma /

epithelioma occurring in people past the climacteric a small ulcer with an indurated base may exist for years, even up to 16 and 20 years without giving rise to more than itching burning and occasional paroxysmal pain. But these cases are rare. Sometimes intolerable pain and itching occur even before the tumour has actually appeared. When it has appeared pain and pruritus become constant. Later when ulceration commences and increases there is usually beside the local symptoms emaciation and all the other signs of malignant cachexia. Intermittent bleeding, incontinence of urine and faeces make their appearance when the ulceration is spreading deeply. There is no attempt at repair or at intermittent arrest of the course. The diffuse form of newgrowth if unattended, usually causes death in about two years owing to the enormous destruction of tissue. If operated on early there may not be a recurrence for a long time.

The histological examination of carcinoma of the vulva reveals the typical interpapillary columns of squamous cells penetrating in the form of irregular masses into the subcutaneous tissues. The typical "pearls" /

"pearls" are found giving the condition the name of "Hornkrebs" (horny cancer.)

The actively inflammatory tissue in the immediate vicinity of the growths and the ring of fibrous tissue found encircling it, are here again the response to the virus on the part of the tissues.

In contradistinction to esthiomène we have in this condition a new type of tissue not found in the healthy human economy. This new tissue does not replace a mass of previously existing fibrous growth but supervenes in a normal tissue which has previously been healthy.

We have seen that some writers have considered esthiomène to be malignant in nature. This view was no doubt due to the fact that esthiomenic tissues may undergo malignant degeneration. This took place in one of our cases where esthiomène clearly existed for some time before the condition took on malignant action. No record of any similar case exists. The possibility that these two cases might have been malignant from the outset is out of question in view of the history and of the physical signs present.

Maggie Lawrence.

Age /

Age 35. Pressworker.

Syphilis since marriage, aet 18; history of vulvar affection two years. On admission a mass of hypertrophied tissue was visible in the position of the right labium minus and the clitoris. It presented all the characters of a syphilitic hyperplasia. Three small shallow ulcerations were present discharging slightly and causing itching and burning sensations for which patient sought relief. One inguinal gland was found enlarged in the right groin. A small hard subcutaneous nodule in the vicinity of the anterior pole of the hypertrophied mass was present on the right labium majus distinct from the hypertrophied area.

Sections of the hypertrophied part, nodule and gland showed typical early squamous cancer.

The parts were removed by operation.

No after history was obtained.

Points of interest are:-

Age of patient: 35

Syphilis at 18.

Vulvar trouble lasted three years.

Condition found: large hypertrophied masses typical /

typical of tertiary syphilis; two areas of ulceration.

In addition to these an outlying hard subcutaneous nodule in the labium majus and an enlarged gland. Early squamous carcinoma.

It is evident that in this case we are dealing with tissues that have for three years been the seats of tertiary syphilitic manifestations and that recently malignant degeneration had occurred in them. In the first case a discharge of fetid odour occurred, in the second there appeared itching, burning and stabbing pain. The patient sought relief as a result of these symptoms. The condition was diagnosed as a tertiary syphilitic manifestation and the microscope revealed early cancer. There can be no doubt that the reported cases of esthiomène, where the duration of the disease was comparatively short, where destruction was very extensive, where the ulceration spread without a tendency to heal where there was no hypertrophy, where the glands were enlarged, where there was much fetor, pus and rapid exhaustion were cases of malignant degeneration supervening on esthiomène. Some authors held these as /

as phagedenic and others held that they were probably malignant from the outset.

Part III.

Elephantiasis.

The traditional belief has until recent times been tacitly accepted that there are two kinds of elephantiasis, one which is caused by the parasite filaria sanguinis hominis, occurs in warm countries, and is usually known as "true" elephantiasis or Elephantiasis Arabum, and another kind which occurs in temperate zones termed "pseudo" or "false" elephantiasis and is mechanically caused by the obstruction of lymphatic vessels from any cause.

In more recent times this belief has been seen to have no foundation. The modern view is that no such distinction between true and false forms can be made, for the essential nature and causation are the same in all forms. Coloured races appear to have a readier disposition to elephantiasic enlargements. Kelly noticed that his negro women patients were more prone to hypertrophies in the vulvar regions than his white.

The view has been held until quite recently that a prolonged oedematous condition was sufficient as sole factor to cause "elephantiasic enlargement" and that /

that passive or active congestion was a sufficient cause to produce scleroma.

Careful microscopic examinations have revealed in all cases of elephantiasis inflammatory processes of an active character with infiltrations around blood and lymph vessels.

The staphylococcus albus, bacillus tetragenous, pneumococcus and sometimes mixed microbial infiltrations have been found. The theories that these findings have produced are the following:-

1. A simple oedema remaining non-infected, ends never in a sclerosis.
2. Oedema predisposes to various local inflammatory processes which in their turn cause sclerosis.

It has been found that in an exceedingly small number of patients afflicted by the so-called true or filarial elephantiasis was the parasite found present in the blood. (Brault, l'Elephantiase chez les Arabes en Algerie. Province Medicale 1908 N. 13)

The mechanical theory of causation is accepted by all modern writers. They hold however that in itself it cannot be the sole cause of elephantiasis. Mechanical /

Mechanical obstruction either in the form of lymphadenitis or extensive lymphangitis is a necessary predisposing factor. Inflammatory infiltrations of a slow chronic insidious nature supervenes and the two causes between them produce an elephantiasic enlargement or as it is more often called "a scleroma." The virus may be specific or non-specific. Among the former infections we have the tuberculous virus which in the 5 cases already mentioned produced enormous hypertrophies without ulceration and where among the oedematous and indurated fibrous tissue tubercle foci and occasional tubercle bacilli were found. Another specific virus may be syphilis. It is sometimes very difficult to distinguish between a "syphilitic elephantiasis" and a true gummatous or granulomatous hypertrophy.

Characters of an elephantiasic enlargement.

The part is uniformly enlarged, so that there is some maintenance of the original shape. The "growth" is firmly elastic but may feel quite hard and wooden to the touch. The surface is irregularly convoluted and may have warty enlargements on it. Sometimes a serous fluid is discharged from it. It is usually pinkish white and the skin may present a scaly thickening in parts. Such ulcerations as may be present are due to injury pressure and friction, for the elephantiasic growth has no spontaneous tendency to ulcerate and break down. The clitoris is most often affected, then the labia majora and finally the labia minora. The whole vulva however may be uniformly and generally enlarged.

The symptoms are due to the local mechanical disturbances caused by the enlargement. An enormously hypertrophied clitoris will obstruct the flow of urine or irritate the urethra causing frequency; or the local deformity may displace the urethral canal and cause retention. The vaginal outlet may be blocked. Inconvenience in sitting and walking may be experienced.

The /

The course of elephantiasis is characterised by excessive chronicity and very slow increase. Accidents to the tumour may occur but apart from these the elephantiasic growth may persist through many years without causing other than mechanical troubles.

Histological features of elephantiasis.

Elephantiasic tissue consists of enormously hypertrophied dermic and hypodermic elements. The epidermis is greatly thickened; its layers retain their normal relationship towards each other. The corneal layer is greatly increased in depth; the epithelial strata show oedema in places. In the dermis we find great oedema of the connective tissue which is increased in amount. The type of the connective tissue is "embryonic." The oedema in places fills the interstices of the connective tissue with a gelatinous looking material. The papillae are much enlarged. In deeper places there are bands of definitely organised fibrous tissue with enlarged venous and lymphatic spaces and a tendency to endarteritis. The tissues generally show lymphstasis with a slow chronic fibrous degeneration going on in parts and foci of more active recent inflammation around lymph vessels.

M.M. Forgue and Massabau give a summary of the histology of elephantiasis in these words:- "There is excessive hyperkeratosis with increase in size of the papillae; the subcutaneous tissue is more or less split up by oedema; the lymphatics are dilated, numerous /

numerous embryonic round cells are disseminated throughout the tissues."

Elephantiasis and Tuberculosis.

M.M. Forgue and Massabau published an article in the Revue de Chirurgie, June 1909 entitled: "l' Elephantiasis tuberculeux a propos d'un cas d'elephantiasis tuberculeux de la Vulve." In this essay the authors refer to a work of Calderoni who collected 51 cases of "false elephantiasis" of the vulva in white women due to various causes, which had been published between 1880 and 1890.

Eight of these were due to tuberculous lymphatic obstruction. C.E. Purslow of Birmingham published the case in the British Medical Journal (27th Oct.1911) of a woman who had had peritoneal tuberculosis of the vulva for some time with tuberculous infiltration of the inguinal and omental lymph glands. The Vulva was uniformly enlarged and presented a warty uneven surface with a watery discharge and without ulceration. In spite of careful examination of the tissue no tuberculous foci were found. The parts presented the histological appearances of elephantiasis. In view of the presence of extensive scars in both groins caused by suppuration of lymph glands we may accept it as a certainty that this was a clear case of /

of elephantiasis caused by lymphstasis and to which the tuberculous virus was added.

In cases such as these it is a matter of the greatest difficulty to find the tuberculous foci and the bacillus of Koch. In the five cases reported of so-called "tuberculous elephantiasis" the foci were found and in some cases the bacillus was stained insitu. In spite of the fact that sometimes the elements of tuberculous disease are undiscoverable one is justified in concluding that hypertrophies with sclerosis in tuberculous patients are due to a mechanical lymph obstruction followed by the tuberculous infiltration of the sclerosed areas.

Cases have been reported of mixed infections, where the lymphatic obstruction was due to syphilis and the local infection to tuberculosis. (Danlos, Annales de Dermatologie 1902). It is not impossible that the primary lymphatic obstruction may sometimes be tuberculous and the local infection be of a mixed septic type.

These rare cases of [illegible] elephantiasis due to tuberculous obstruction of the lymphatic system differ from esthiomène in the following points:-

1. The /

1. The enlargement is usually uniform, a hyperplasia without loss of shape, often the whole vulva is uniformly enlarged. In esthiomenic enlargement however one or other part is enlarged and totally altered in shape and appearance. An inspection of Huguier's excellent plates of 6 cases will point out this characteristic.
2. In this form of elephantiasis there is usually no ulceration. If ulceration occurs it resembles the ulcers obtained in cases of elephantiasis from various causes. They are trophic or traumatic, chiefly the latter, due to pressure or friction. In esthiomene on the other hand the ulceration is definitely active in character with a tendency to extend downwards and inwards, canalising the tissues it attacks.
3. The history usually helps in pointing out the nature of the disease; there is almost always phthisis present or tuberculous glands in the neck and sometimes tuberculous peritonitis.
4. Scars with enlarged glands in the groin may be present in esthiomene and in elephantiasis due to lymphatic obstruction. The appearance of the /

the scars, their history and duration, and the feel of the glands will teach their origin. If they are syphilitic there may be similar scars elsewhere or another skin condition may assist in forming a diagnosis. The history of the case will often be sufficient in indicating whether the scar is due to lupus or to some previously discharging tuberculous gland

Elephantiasis and Syphilis.

Syphilis is a frequent cause of lymphatic obstruction. The spirochaeta pallida is known to make its progress into the blood stream by the lymph channels. In the primary stage small lymph vessels are sometimes to be seen crammed full of organisms, The lymph gland enlargement is one of the first manifestations of the disease. Formerly the "mixed infection" with suppuration and slow healing of the bubo was very common. The typical scar resulting was always looked for. These septic infections are not so often seen in our days. Lymphangitis and lymphadenitis due to syphilis cause an indurative state of these structures which if extensive produce lymphstasis, and a chronic oedematous state. This chronic oedematous state is often seen in the vulva. For a long time it may remain merely as an oedematous local enlargement. This has been made known as "syphilitic elephantiasis" by many writers.

Sooner or later however the unhealthy indurative tissue becomes the seat of some pathological agent which may be any microorganism. If it is one or several of the ordinary septic microbes, commonly found /

found on unhealthy skin surfaces, a septic ulceration sooner or later takes place; it is usually an indolent chronic slowly spreading and slowly healing sore which may discharge pus. Sometimes the bacillus of Koch has supervened, but that is very rare. (Danlos) The onset of the ulceration may be determined by the same organism which caused the lymphstasis, namely the spirochaeta pallida.

Elephantiasis and Esthiomène.

From the foregoing it will be seen how difficult it is to distinguish between a "syphilitic elephantiasis" and esthiomène.

All syphilitic enlargements in the region of the vulva are not elephantiasic, i.e. oedematous enlargements due to lymphstasis. A large number of them are

a. gummata.

b. a massive infiltration of one or more parts of the vulva which may become generalised over the whole of its extent (Prof.Fournier)

Both these tertiary syphilitic manifestations differ from the elephantiasic enlargement by being typical granulomatous tissue containing the spirochaeta pallida and showing all the characteristics of tertiary syphilitic tissue, while in the latter condition we have normal though unhealthy oedematous tissue presenting the characters seen in other cases of elephantiasis.

We wish to draw a definite line of demarcation between such cases of hypertrophy that are due to an oedema caused by syphilitic lymph obstruction and such /

such cases of hypertrophy which arise directly from the action of the syphilitic virus on the tissues. All the latter we unite under the heading of esthiomène, using a long established term conveniently instead of the somewhat cumbrous expression of "syphilitic hypertrophy with ulceration." Where the hypertrophy is clearly due to lymphatic obstruction, where it is uniform, oedematous and presents no ulceration or only a very slight form the expression of elephantiasis should be retained. The majority of cases published as "syphilitic elephantiasis" (Veit, Kelly) have such characters as to place them into the field of esthiomène. Where the beginnings of the condition have not been seen it may be extremely difficult to distinguish between cases of esthiomène and of elephantiasis; but there is almost always some feature which will help to form an exact diagnosis, the microscope will show granulomatous tissue in one case, and loose embryonic oedematous dermic and hypodermic structures in the other.

The main points which distinguish an elephantiasic enlargement from esthiomène are the following:-

1. Extensive lymph gland infiltration with scars in the /

the inguinal regions, denoting a great degree of lymphatic obstruction determines elephantiasis. Esthiomène is not dependent on this fact. Often no enlargement of glands and no scars are found in the groin yet we have extensive hypertrophy in one part or other of the vulva.

2. In elephantiasis if the term is correctly applied the enlargement is uniform and the original shape of the part is maintained. In esthiomène however it is irregular and the masses of heaped up tissue assume the weirdest and most extra-ordinary forms. The original parts are no longer recognisable.

3. A histological examination reveals eodematous tissue with dilated lymph spaces and occasional inflammatory groups of cells in elephantiasic tissue. We have discussed the histology of esthiomène at some length and recognised its gummatous nature, its granulation tissue, the active inflammatory reaction to the syphilitic virus in the tissues.

4. Course and result.

Elephantiasis is more chronic than the most chronic /

chronic of all cases of esthiomène. It may remain the same for years since it does not break down. It causes no disturbance of health and does not lead to death. It is merely a disturbing result of insufficient lymph circulation producing an unhealthy state of the vulva. On the other hand esthiomène is caused by the syphilitic virus in its tertiary or attenuated form; it is capable of producing the most profound constitutional disturbances early in its course, with later cachexia and death.

5. Ulceration in syphilitic elephantiasis is very slow chronic and limited. In cases of esthiomène it tends to be present to an equal degree with hypertrophy and in later and severe cases is greatly in excess. The destruction of organs is sometimes immense. Cases are on record where the whole perineum was eaten away and the pelvic organs had become a scarcely to be differentiated mass of ulcerating tissue.

Part IV.

Kraurosis Vulvae.

This is a condition which as Jayle and Bender, Berkeley and Bonney have pointed out is not a disease per se, but is sometimes the atrophic result of a long continued chronic vulvar inflammation, and sometimes the manifestation of " ovarian insufficiency." The nature of the previous inflammation may be simple or syphilitic. That it is often an indication of " ovarian insufficiency " is shown that by the fact that it occurs in

1. Young sterile women
2. After premature menopause.
3. After double oophorectomy.

Its essential nature is an equal gradual shrinking of all the skin layers and subcutaneous tissues with no new histological features added. The labia majora and minora, the clitoris and vestibule are simultaneously affected. In the later stages there is narrowing of the urethral and vaginal outlets. Pain and soreness accompany the shrinking process until the atrophy is complete. According to Berkeley and Bonney the process is not unlike physiological senile atrophy but more rapid and complete.

Leukoplakia.

Like leukoplakia of the tongue this condition when occurring on the vulva has been held by some observers to be invariably due to syphilis. Berkeley and Bonney, however hold that the chronic inflammatory process which initiates it, is of unknown origin. There can be no doubt that syphilitic infection has taken place in many cases of leukoplakia of the vulva.

Berkeley and Bonney distinguish four stages:

1. The affected parts are red, swollen, dry and excoriated.

   The surface epithelium desquamates and there is diffuse subepithelial lymphocytosis.

2. Retraction takes place in some parts and thickening in others; some patches show a white semi-opacity.

   A hyaline zone of de-elasticised tissue under the surface shows atrophy of the interpapillary columns of epithelial cells. Pruritis is present during both these stages.

3. Cracks and ulcers occur; bleeding is common, also pain owing to exposure of nerve-endings.

Carcinoma /

Carcinoma frequently supervenes at this stage. Keratinisation of the surface epithelium is accompanied by general sclerosis of the sub-epithelial tissues which show complete atrophy of the interpapillary epithelium.

4. General atrophy and quiescence of vulva. The disease may spread to thighs and perineum.

. Complete keratinisation and sclerosis of the tissues.

Kraurosis, Leukoplakia and Esthiomène.

It is not a matter of difficulty to distinguish conditions so different from esthiomène as kraurosis and leukoplakia. These conditions are as we have seen the results of local chronic inflammation with the exception of those cases that are due to insufficient ovarian secretion (kraurosis) and those that are a manifestation of tertiary syphilis (leukoplakia). The majority of the cases of leukoplakia and kraurosis are local atrophic degenerations after some long-continued irritation, usually inflammatory. Esthiomène is always a manifestation of tertiary syphilis.

The essential changes in kraurosis and leukoplakia both macroscopic and microscopic are atrophic changes. Ulcerations if they occur (leukoplakia) are purely accidental. In esthiomène the main change is a hypertrophy followed by a process of necrosis.

There are some local disturbances in kraurosis and leukoplakia, pain and general soreness in the former, and intolerable pruritis, dysuria and pain in the latter. We have seen that esthiomène may exist /

exist for a very long time without giving rise to trouble. The result of a severe case of esthiomène is exhaustion and death. In leukoplakia and kraurosis the process wears itself out without producing such dire results. Carcinoma may supervene on leukoplakia (Berkeley and Bonney) but leukoplakia without carcinoma is never fatal.

The treatment by medication is not attended by any results in leukoplakia and kraurosis even when they are due to syphilis. Complete excision of the parts is the only procedure which is likely to shorten the long and tedious course of these conditions. Many cases of esthiomène undergo a rapid healing under the influence of mercurials combined with potassium iodide.

CHAPTER VIII.

Treatment.

The lines of treatment are the following:-

1. General, or constitutional and
2. Local.

If possible the patient should rest in bed until all ulcerations are healed. In severe cases and very poor patients a prolonged stay in a hospital should be enforced. The object is to prevent all further irritationof the parts, to enforce absolute cleanliness and the carrying out of consecutive treatment and to improve the general health by regular and ample feeding. With rest, cleanliness feeding and medicinal treatment esthiomène frequently disappears without returning in cases of recent occurrence and comparatively acute character.

The main medicinal treatment consists in giving mercurials combined with potassium iodide. Sometimes the former have more influence than the latter and it is a wise plan to carry the mercurial treatment up to the point of soreness of gums and slight salivation.

There is no need here to enter into the details of /

of the treatment of syphilis by mercurials. It suffices to say that the more recent the case the more likely it is that the condition yields to treatment. When the condition is of some years standing there is less likelihood of cure by internal remedies. Potassium iodide should in all cases be added to the mercurials and be pushed to large doses.

Locally strict cleanliness must be observed. Frequent warm baths are indicated. The parts should be bathed with soothing lotions several times; if there is much itching, a lotion containing a little opium keeps the parts quiescent and prevents scratching and rubbing. Lotio nigra should be freely used and the parts then dried and dusted over with a mixture of boracic and calomel powder. If there is incontinence of urine and faeces special care and trouble should be taken to keep the perineum as clean and dry as possible; this is sometimes an impossibility. It is a good plan to touch ulcers with a 5% solution of Silver Nitrate.

Operative measures are frequently indicated and prove of great value in many cases where the disease has shown no inclination to respond to treatment, where /

the hypertrophy goes on increasing and where the ulceration assumes more and more a burrowing character.

Operative treatment consists in removing the hypertrophied masses en bloc. This sometimes necessitates the removal of large masses of tissue representing hypertrophied structures of the vulva. If there is much bleeding only a few cuts with the knife should be made at one time and the remaining raw surfaces brought in apposition by a few sutures ligatured tightly to stop the bleeding; then another small portion should be severed from the base and the surface ligatured and so on until the whole mass is removed. Where superficial callous ulcerations exist they are often benefited by scraping with a sharp spoon and freshening up their edges. Skingrafting is not likely to have good results. When the ulcerations are deep and widespreading nothing can be done surgically. Fistulae cannot be cured with operation on account of the unsoundness of neighbouring tissues through which stiches would simply tear. Strictures after healing require gradual dilatation.

Surgical treatment must not replace medicinal. Antisyphilitic remedies must be persevered with even if /

if the operative measures have been successful in promoting healing. It must always be remembered that recurrence of esthiomène may take place in which case the treatment in all its details must be resumed.

Finally we wish to indicate the great importance of early removal of tissue for purposes of microscopic examination. This may reveal an early cancerous degeneration as in the case of Maggie Lawrence and the early removal of the affected parts and glands in the groin may save the patients life for many years.

In a case of permanent cure when owing to great ulcerative destruction and cicatricial contraction and operative interference great deformity of the vulva vagina and urethra has resulted, much can be done to restore these by extensive plastic operations. These are however not indicated in those cases where some ulceration still persists and where the cure is of recent date.

SUMMARY AND CONCLUSION.

I.

Nomenclature.

The term esthiomène, established in medical literature by the highest authorities of the 19th century has been misunderstood and misapplied by many authors.

We hold that it should be retained as a useful term replacing the expression "hypertrophy with ulceration." It should be clearly understood to mean a tertiary syphilitic manifestation.

"Lupus Vulvae" should be replaced by the expression "tuberculosis of the vulva." Lupus vulgaris as it presents itself on the skin has not been observed in the region of the perineum. Tuberculous ulcerations and tuberculous hypertrophies however do occur.

The term "elephantiasis" has been applied to hypertrophies occurring in chronically oedematous parts whence the return of lymph has become obstructed or rendered sluggish and where owing to the unhealthy state of the chronically enlarged parts a low /

low form of chronic inflammation has set up. The cause of the oedema and the micro organisms which determine the onset of these inflammations differ widely. Any virus may cause it. Sometimes it is due to syphilis. Hence with regard to cases of elephantiasis caused by the spirochaeta pallida we hold that the name should be carefully applied only to those which show evident lymph obstruction and oedema, where the hypertrophy is extensive and where there is little or no ulceration. In esthiomène, which is always syphilitic, there is no oedema, no evident lymph obstruction; the hypertrophy and ulceration may be equal in degree but in severe cases the latter is very extensive. Esthiomène is not an oedema but a granulomatous growth with a tendency to necrosis.

II.

The Nature of Esthiomène.

1. Esthiomène is not a disease sui generis.
2. It is not a form of low chronic ulceration occurring on a soil weakened by constitutional syphilis or tuberculosis.
3. Esthiomène is not merely a local inflammatory state following on irritation.
4. There is no relationship between tuberculosis and esthiomène.
5. The only connection there is between esthiomène and malignant disease is, that the former may, very occasionally undergo malignant degeneration.
6. Esthiomène is not due to lymph stasis hence it does not belong to the group of hypertrophies commonly called "elephantiasic."

III.

Esthiomene a tertiary syphilitic manifestation.

a. A probable or direct history of syphilitic infection is always obtainable.

b. The majority of early cases of esthiomène respond to antisyphilitic treatment. The late and chronic cases which do not respond do not therefore indicate that sometimes esthiomène may be due to other causes; there are other tertiary lesions such as stricture of the rectum, leukoplakia of the tongue and of the vulva which are equally resistant to antisyphilitic treatment.

c. The slow chronic course of esthiomène marked by attempts at healing with subsequent relapses, the absence of local disturbances, the non-impairment of general health indicate the syphilitic nature of the condition.

d. The masses of cicatricial tissue with subsequent contraction producing severe strictures and extensive deformities which result from a syphilitic lesion when the cicatrising process is in excess over the ulceration is typical of no other /

other disease. When on the other hand the necrosis exceeds the tendency to fibrosis we get enormous tissue destruction with profound cachexia and death.

e. In no other constitutional disease is there such a constantly present combination of hypertrophy and ulceration as in syphilis. Where hypertrophies are extensive there may be some ulceration or there may be none; this is obtained in elephantiasis. Where ulceration is extensive, such as in tuberculosis there is usually little or no hypertrophy.

f. The microscope reveals the typical gumma or granuloma of the third stage.

g. We find no case recorded where the spirochaeta pallida has been found in the tissues affected by esthiomène.

It is a well-known fact that the organism does exist, though in comparatively small numbers in the lesions of the tertiary stage, but they are usually difficult to find (D'Arcy Power and McKeoch, System of Syphilis). With the easier methods of demonstration of the organism, the time /

time will no doubt come when the spirochaeta of Schandium will be shown in esthiomenic tissues with the same ease as the bacillus of Koch in tuberculous lesions.

h. With equal certainty of proof will the positive Wassermann reaction relegate all cases of esthiomene into the field of tertiary syphilitic lesions.

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